

Sixth Edition

Short Cases in **SURGERY**



S.K. Bhattacharya

VISIT...

LANZAROTE
Caliente.COM

Short Cases in **SURGERY**

SIXTH EDITION

Swaraj Kumar Bhattacharya

M.S. (Cal.), F.R.C.S. (Edin.)



CBS PUBLISHERS & DISTRIBUTORS PVT. LTD.

NEW DELHI • BENGALURU • PUNE • KOCHI • CHENNAI

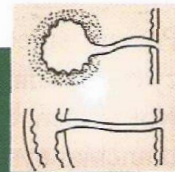
CONTENTS

Contents

<i>Preface to the Sixth Edition</i>	vii
<i>Preface to the First Edition</i>	ix
1. Lumps, Ulcers and Fistula/Sinus	1
2. Skin and Subcutaneous Tissue	16
3. Swellings in the Neck	75
4. Cervical Lymphadenopathy	103
5. Salivary Glands	117
6. Swellings of the Jaw	131
7. Actinomycosis	140
8. The Mouth and the Tongue	143
9. Umbilicus and Abdominal Wall	157
10. Hernia	163
11. Testis, Epididymis and Scrotum	196
12. Penis	219
13. Filariasis and its Surgical Manifestations	229
14. Raynaud's Syndrome (Syn. Raynaud's Phenomenon)	234
15. Aneurysm	244
16. Varicose Veins	254
17. Ulcers of the Leg	265
18. Swellings of the Leg	272
19. Congenital Anomalies	280
20. Soft Tissue Sarcoma	300
21. Intestinal Stomas	309
22. Orthopaedic Disorders	320
<i>Index</i>	339

1

CHAPTER



Lumps, Ulcers and Fistula/Sinus

CLINICAL EXAMINATION OF A LUMP OR SWELLING

This should be conducted in a systematic manner – history, local examination, examination of regional lymph nodes, and general examination.

HISTORY

- Age, sex, ethnic group, occupation.
- Duration: *How long the lump is there.*
- Mode of onset: *How the lump has started.*
- Progress of the swelling: *Is the lump changing its size and surface.*
- Painful or painless?
- Family history.

LOCAL EXAMINATION

Inspection

- Site
- Number
- Shape and size
- Colour
- Surface and edge
- Skin over the swelling
- Visible pulsation
- Impulse on coughing

Palpation

- Local temperature
- Tenderness
- Site, size and shape

- Surface and edge
- Consistence
- Fluctuation
- Translucency
- Fluid thrill
- Resonance
- Compressibility
- Pulsatility
- Indentation
- Impulse on coughing: Visible or palpable.
- Reducibility
- Mobility

Solid
or
Fluid
or
Gas

Vascular

Percussion

Auscultation

STATE OF LOCAL TISSUES

STATE OF REGIONAL LYMPH NODES

GENERAL EXAMINATION OF THE PATIENT

LUMP

A lump or swelling may arise from the skin, subcutaneous tissue, muscle, tendon, bone, nerve, lymphatics and lymph nodes, blood vessels, gland, or lie within one of the body cavities.

A lump may also arise due to protrusion of a viscus from the body cavity through a weak spot of its wall, e.g., hernia.

Causes of a lump or swelling

A. Congenital

- Present since birth, e.g., meningocele, dermoid cyst, teratoma.
- Not evident at birth but appears years later, e.g., branchial cyst, thyroglossal cyst, cystic hygroma.

B. Acquired

- Traumatic: Swelling develops immediately after a trauma, e.g., haematoma, muscular lump caused by rupture, bony projections following fracture or dislocation.
- Inflammatory: Acute or chronic.
- Neoplastic: Benign or malignant.
- Otherwise, e.g., autoimmune disease.

DIAGNOSIS OF A LUMP OR SWELLING

History

- Age, sex, ethnic origin, occupation.
- Duration: *How long the lump is there?*
 - Congenital and present since birth, e.g., meningocele, dermoid cyst, teratoma.
 - Congenital but not evident at birth and appears years later, e.g., branchial cyst, thyroglossal cyst, cystic hygroma.
 - Acquired.
- Mode of onset: *How the lump has started?*
 - The swelling may appear after trauma or develop spontaneously. A swelling may arise on some pre-existing condition, e.g., a keloid may develop over a scar; malignant melanoma may develop over a mole or birth mark.
- Progress of the swelling: *Whether the lump is changing in size or surface.*
 - Rapid enlargement may imply inflammation particularly if it is painful, whereas progressive enlargement may signify neoplasia.
- Painful or painless?
 - Painful lumps are commonly due to trauma and inflammation. Pain is absent in benign growth and in early carcinoma.

Local examination

Inspection

1. *Site:* Location of the lump and its relation to certain landmarks present in the vicinity of the lump.

Typical position of a swelling is almost diagnostic of some lesion, e.g.

- Post auricular dermoid behind the ear;
 - External angular dermoid at the lateral end of the eyebrow;
 - Meningocele over the back in the midline;
 - Thyroid swelling in front of the neck.
2. *Number:* Whether single, or multiple, e.g., neurofibromatosis, adiposis dolorosa, warts, naevi, secondary carcinomatous nodules, diaphyseal aclasia.
 3. *Shape and size:* Whether spherical, ovoid, kidney-shaped, pyriform or irregular. The lump may be pedunculated or sessile.
 4. *Surface:* Smooth (cyst), lobulated (lipoma), nodular (a mass of enlarged lymph nodes, multinodular goitre), rough and irregular (irregularity of a wart; carcinoma), red, shiny and oedematous (inflammatory lesion).

There may be presence of a *pit or punctum* (sebaceous cyst), *peau d'orange* (due to underlying carcinoma infiltrating towards the skin), *scar* (irregular scar from injury; broad, depressed and puckered scar from suppuration; linear scar with suture marks from previous operation – *this indicates that the lump appears after operation, e.g., incisional hernia; recurrence of lump, e.g., Paget's recurrent fibroid. Tense shiny skin with overlying prominent veins* (sarcoma).

There may be *ulcer over a swelling* (necrosis over a malignant nodule).

5. *Edge of the swelling:* Whether circumscribed or of the nature of a general bulge, i.e., without any definite margin. The edge may be sharp, rounded, regular or irregular.

Well-defined and regular (most of the benign swellings); *well-defined and irregular* (malignant swelling infiltrating the surrounding skin); *diffuse and ill-defined* (inflammatory swellings like cellulitis, abscess).
6. *Colour:* Red or purple (inflammation, haemangioma), blue (ranula, mucous cyst), black (melanoma).
7. *Visible pulsation:* Visible expansion synchronous with each pulse beat (aneurysm, vascular growth).

8. *Visible impulse on coughing* depending on the site of the lump. When the swelling is noticed over the groin, abdomen, chest, spine or cranium: *Note cough impulse*, e.g., hernia, meningocele.
9. *Movements of the swelling on deglutition* when the swelling is situated in front of the neck – thyroid swelling moves up on deglutition.
10. *Note any pressure effects* when the swelling is on the limb – inspect the distal part for any pressure effect like oedema, foot drop, wrist drop from nerve palsy.

Palpation

Before starting palpation of the lesion the patient should be asked whether the lesion is painful and/or tender.

1. *Temperature*: Raised in presence of acute inflammation, cellulitis or abscess. Some vascular tumours (e.g., sarcoma) may also show raised temperature.
2. *Tenderness (pain on pressure)*: Present over a traumatic or inflammatory lump and is a feature of inflammation. Tenderness is also often noted over a rapidly expanding malignant lump. A swelling along the nerve is also tender, e.g., solitary neurofibroma. A swelling caused by bony fracture is also tender.

Benign growth is painless and non-tender.

3. *Confirm the site, size and shape, surface and edge*. Edge may be well-defined or ill-defined. *In lipoma the edge is well-defined and when pressed upon it slips away under the examining finger (slipping sign) – this is characteristic of lipoma.*

Measure the size of the swelling with tape in 3 dimensions – length, breadth and height.

4. *Consistence*: Depends on the contents of the lump which can be composed of cells, fluid or gas. So, consistence varies from soft to bony hard – *soft* (lipoma, haemangioma), *cystic* (cyst, aneurysm and abscess), *firm* (fibroma, papilloma), *hard but yielding* (chondroma), *stony hard* (metastatic lymph nodes), or *bony hard* (osteoma, osteochondroma).

Consistence may be uniform throughout or variable. *Variegated consistency* i.e., some part is soft, some part firm, some part hard – suggests malignant growth.

A cystic swelling feels softer at the centre than at the periphery. *A solid swelling* feels firm at the centre than at the periphery.

5. *Indentation or moulding*: Certain cysts like sebaceous cyst or dermoid cyst contain pultaceous or putty-like material and can be moulded. Such swelling when pressed upon with the tip of the finger, in contradistinction to the sign of emptying, it stays indented. Such sign of indentation can also be demonstrated over the palpable sigmoid colon loaded with solid faeces, in the left iliac fossa.
6. *Compressibility*: Some soft or cystic lumps are compressible. When they are squeezed they diminish in size considerably or disappear, but reappear slowly on release of pressure. This is known as *the sign of emptying* and is characteristic of blood-filled lesions such as haemangioma, aneurysm but may also be seen in lymphangioma and narrow-necked meningocele.

The sign of emptying in haemangioma may be accompanied by blanching, i.e., the lesion becomes colourless or pale as the blood is expressed out of the lesion. The colour returns on release of pressure.

7. *Fluctuation*: This determines the presence of fluid (or gas) in a swelling – *cystic swelling*. This clinical sign can be elicited in a non-tense cystic swelling where an impulse transmitted to the finger or fingers of one hand on one side of the swelling by sudden pressure exerted with the finger or fingers of the other hand on the other side of the swelling and vice versa. Fluctuation implies transmitted impulse in two planes at right angles to each other. *So, fluctuation should be elicited in both directions right angles to each other*, as fleshy muscle in the thigh can be fluctuant across but not in longitudinal directions.

Fluctuation is positive in cystic swellings like hydrocele, cystic hygroma, meningocele, bursa, etc. For small swellings (< 2 cm) do the Paget's test.

Pseudo or false fluctuation is felt in a large soft swelling but without any fluid content e.g., lipoma.

Cross fluctuation is fluctuation between two separate swellings but communicating with each other e.g., *compound palmer ganglion in the wrist* above and below the flexor retinaculum; *psoas abscess pointing to the thigh* above and below the inguinal ligament; *deep plunging ranula in the floor of the mouth* extending to submental region.

8. *Transillumination*: Once the swelling is found to be cystic, *test for translucency* in order to determine the nature of the fluid in the cyst – clear or otherwise. *Cystic swelling containing clear fluid* (e.g. hydrocele, meningocele, cystic hygroma, ranula, bursa, etc.) *will transilluminate with pointed light provided the covering skin is thin.*

Hydrocele due to filaria, haematocele, though cystic, will not transilluminate.

Brilliantly transilluminable swellings:

- Cystic hygroma
- Congenital hydrocele
- Epididymal cyst
- Ranula
- Meningocele with thin skin cover.

For any scrotal swelling transillumination test is a must. Any scrotal swelling when felt hard and transillumination is negative: consider testicular tumour and proceed accordingly.

9. *Pulsatility*: A swelling may pulsate synchronous with each pulse beat of the patient.

The pulsation may be *expansile pulsation* where the swelling expands in all directions as well as it pulsates, e.g., aneurysm, vascular malformation, arteriovenous fistula.

This pulsation of the swelling may be *transmitted pulsation* where the swelling does not increase in size but is merely raised with each throb of the underlying artery, e.g., a pancreatic mass situated in front of the abdominal aorta, a carotid body tumour at the bifurcation of the common carotid artery.

A carotid body tumour may have rich blood supply and may exhibit expansile as well as transmitted pulsation. Some other tumour like telangiectatic type of osteogenic sarcoma have a rich blood supply to show expansile pulsation.

In case of an aneurysm, pressure over the main artery proximal to the swelling results in

diminution in the size of the swelling as well as pulsation.

10. *Reducibility*: It indicates that a swelling can be emptied by squeezing but the swelling does not return spontaneously on release of pressure (cf. compressibility) – reappearance of the swelling requires an additional force, e.g., straining or coughing, or the effect of gravity. *A classic example is hernia.*
11. *Impulse on coughing*: This is present in those swellings which are soft or cystic and are likely to be in continuity with the interior of one of the body cavities, i.e., abdomen (external hernias, iliopsoas and lumbar abscesses), chest (e.g., empyema necessitatis), spine and cranium (e.g., meningocele). *The impulse on coughing is usually both visible and palpable and when present, say cough impulse is positive.*
12. *Thrill on palpation*: This is commonly found in an arteriovenous fistula. A thrill may also be present in toxic goitre.
13. *Mobility of the lump*: To know the depth i.e., anatomical plane the lump is situated and its relations to the surrounding structures, i.e., to skin, subcutaneous tissue, fascia, muscle, tendon, bone, vessels and nerves. The mobility of a lump depends on its site of origin as well as on its tethering and fixation to the surrounding tissues. A lump may be attached, adherent or fixed to the skin, subcutaneous tissue, deep fascia, muscle, tendon, bone, vessels or nerve.
- (a) *Fixity to the skin*: A lesion arising from the skin e.g., papilloma, wart, mole, sebaceous cyst, keloid, epithelioma etc., moves with the movement of the skin.

Swelling deeper to the skin: Try to pinch the skin over the swelling at various places or try to move the skin over the swelling by sliding movement:

Lipoma – skin can be pinched suggesting the swelling is not fixed to the skin.

Malignant breast lump – skin cannot be pinched or slid over the swelling as the swelling is fixed to the skin. Even if skin can be pinched over the swelling, move the skin or the swelling – the skin can be seen puckered as seen in early carcinoma of

breast when the swelling is said to be tethered to the skin. *Tethering* is indirect fixity to the skin and is due to fixity of fibrous septae due to infiltration e.g., infiltration of Cooper's ligament in carcinoma breast.

- (b) *Subcutaneous lump* is free from the skin and moves freely over the contracted muscle. *Contraction of the underlying muscle and fascia make a subcutaneous lump more prominent and easily palpable.*
- (c) *Fixity to the deep fascia* only is difficult to demonstrate. *A lump beneath the deep investing fascia of the neck becomes less palpable by pressing the chin on the opposite side of the swelling against the examiner's hand.*
- (d) *Fixity to the muscle:* A lump may be attached to a muscle, or incorporated in the muscle or situated underneath the muscle.

When the muscle is relaxed the lump may be easily palpable and freely movable across the long axis of the muscle.

When the muscle is contracted, the swelling may change its character:

- The swelling may become prominent as when it lies above the muscle.
- The swelling may remain unaltered as when it is incorporated in the muscle.
- The swelling may diminish in size or impalpable as when it lies under the muscle.

Mobility of the lump over the relaxed and contracted muscle – a lump which is adherent to the muscle becomes immobile when the muscle contracts.

- (e) *Fixity to the tendons:* A swelling in connection with the tendon of a muscle becomes fixed when the concerned muscle is contracted against resistance. A swelling in connection with a tendon also moves with the tendon on movement of the concerned muscle.
- (f) *Fixity to the bone:* Swelling in connection with the bone cannot be moved apart from the bone e.g., jaw tumour, exostosis.

- (g) *Fixity to the vessels and nerves:* A lump in connection with a vessel or nerve **can be moved across** (i.e., at right angles to their axes) but not along the direction of their axes.

Percussion

To differentiate gaseous swelling from fluid or solid swellings, the first being resonant, the latter two are dull. Tympanic or resonant in enterocele or pharyngocele. In a small swelling percussion is not done.

Auscultation

All pulsatile swellings should be auscultated with a stethoscope for any bruit or murmur. It may be systolic, diastolic or continuous murmur. A continuous buzzing sound, known as machinery murmur or bruit is audible over an arteriovenous fistula.

Bruit is also heard over a very vascular lesion e.g., thyrotoxic goitre.

State of the local tissues

1. *Presence of induration:* Induration implies thickening and firmness of the surrounding tissues and is due to inflammatory oedema or infiltrating neoplasia. The indurated area may be indented or depressed on pressure, or hard. It may be tender.
2. There may be ecchymoses, pigmentation, metastatic nodules, prominent vessels.
3. If the swelling is overlying a bone – feel for bony indentation or erosion deep to the margin of the swelling e.g., dermoid cyst, meningocele.
4. If the swelling is on the limb
 - (i) Look for oedema, dilatation of veins due to pressure over the vein.
 - (ii) Palpate the distal pulse for any pressure over the artery.
 - (iii) Look for wasting of muscles, paresis or paraplegia, foot drop, wrist drop due to pressure over the nerve.
5. Examine the neighbouring joints, above and below the swelling to note whether there is impairment of joint movements.

State of the regional lymph nodes

The draining lymph nodes must always be **palpated** for involvement in presence of inflammatory or malignant lesion.

The skin, muscles and bones of the limbs and trunk drain to the axillary and inguinal nodes.

The head and neck drain to the cervical nodes.

The intraabdominal structures and testes drain to the pre- and paraaortic nodes.

General examination

Look for any systemic effects produced by infection, trauma or malignancy.

Is there similar swelling elsewhere, e.g., neurofibromatosis, diaphyseal aclasia.

In case of suspected malignant growth a search should be made for secondary deposits elsewhere, e.g., *neck, axillae and groins* for enlarged lymph nodes; *abdomen* for enlarged, hard and nodular liver, mass due to enlarged nodes, ascites; *chest* for pleural effusion or consolidation.

DIAGNOSIS OF A CYSTIC SWELLING

1. **Site:** Most of the congenital cystic swelling has a typical location such as follows:
 - *Branchial cyst* – anterior triangle of the neck, partly covered by upper one third of sternomastoid.
 - *Dermoid cyst* – along the planes of fusion such as midline of the body e.g., at the root of the nose; on the scalp e.g., peteryon, asteryon (post auricular dermoid), on the inner or outer angles of the eye (angular dermoid).
 - *Menigocele* – in the new born along the mid line of the back, commonly at lumbosacral region.
 - *Ganglion* – on the dorsum of the hand.
2. **Shape:** Majority cystic swellings are round and oval
 - *Subhyoid bursitis* – Transverse oval cystic swelling in the midline of the neck.
 - *Thyroglossal cyst* – Vertical oval cystic swelling in the midline of the neck.
 - *Sebaceous cyst, dermoid cyst* – globular or hemispherical swelling.
3. **Surface:** Most cystic swellings in the skin and subcutaneous tissue have smooth surface.
4. **Consistence:** Cystic or firm to hard. *Fluctuation is positive in all cystic swellings.* Fluctuation is to be elicited in two directions right angles to each other.

- *Soft cystic swelling* – Menigocele, thyroglossal cyst, cystic hygroma, aneurysm.
- *Tense cystic swelling* – Hydrocele, ganglion. A tense ganglion may feel hard.
- *Soft cystic at the centre with firm thickened periphery* – cold abscess.

- *Putty or tooth paste* – Sebaceous cyst, dermoid – the swelling is usually indented by a finger tip.

5. **Signs of compressibility:** The swelling may disappear completely or partially on application of pressure, but reappears on release of pressure e.g., aneurysm, haemangioma, lymphangioma, meningocele.

6. **Pulsations:** Expansile or transmitted.

- *Expansile pulsation* – aneurysm.
- *Transmitted pulsation* –
 - (i) when a swelling lies over a vessel e.g., pseudopancreatic cyst in front of abdominal aorta;
 - (ii) when an artery is pushed by a structure underneath e.g., subclavian artery over the cervical rib.

7. **Transillumination:**

- *Positive* with cysts containing clear fluid e.g. hydrocele, meningocele, cystic hygroma.
- *Negative* with cysts containing thick or opaque fluid e.g. hydrocele due to filaria, haematocele.

ULCERS

An ulcer is a persistent breach in continuity of the surface epithelium – the skin or mucous membrane. This is due to a gradual necrosis of the surface tissue which results in the formation of a sore or ulcer. The dead tissue, which becomes separated from the living tissue during the process of necrosis, is termed as slough.

Causes of ulcer (or classification)

- Non-specific
- Specific
- Malignant

A. Non-specific

1. Mechanical
 - Traumatic – Dental ulcer (caused by sharp tooth or ill-fitting dentures), pressure sores (caused by prolonged pressure, e.g., by splints or plasters, or prolonged bed rest).

- Physical – Burns and scalds, electric burn, X-ray burn.
- Chemical – Caused by acids or alkalis.
- 2. Infective: Due to secondary infection of wounds by non-specific pyogenic organisms like *Staphylococcus* or *Streptococcus* (pyogenic ulcer, Bairnsdale ulcer).
- 3. Trophic or nutritional (Greek *Trophe* = nutrition) – due to an impairment of the nutrition of tissues which depends upon adequate blood supply and nerve supply.
 - Vascular insufficiency: *Usually causes painful ulcers.*
 - Poor arterial supply: Burger's disease, Raynaud's disease, arteriosclerosis.
 - Venous stasis: Varicose ulcer, venous ulcer in post-phlebitic limb.
 - Neuropathic: *Usually causes painless ulcer.* Ulceration is due to sensory impairment or anaesthesia, e.g., tabes dorsalis, syringomyelia, leprosy, diabetes, spina bifida, peripheral nerve injury. *Neuropathic ulcers are often called perforating ulcers.*
- 4. Tropical ulcer.
- 5. Cryopathic ulcer.
- 6. Martorell's ulcer (hypertensive ulcer).
- 7. Bazin's ulcer (erythrocyanoid ulcer).

B. Specific

Due to specific infections

1. Tubercular ulcer
2. Syphilitic ulcer
3. Soft chancres
4. Leprosy
5. Actinomycotic ulcer
6. Meleney's ulcer.

C. Malignant

1. Rodent ulcer
2. Epithelioma
3. Malignant melanoma
4. Any malignant growth fungating through skin.

CLINICAL EXAMINATION OF AN ULCER

This should be conducted in a systematic manner – history, local examination, examination of regional nodes, general examination.

History

The following points to be enquired:

1. Age, sex, ethnic origin and occupation
2. Mode of onset

How has the ulcer developed?

- Following trauma – traumatic ulcer.

OR

- Spontaneously:

- over a nodule or lump, e.g., tuberculous lymphadenitis or abscess, gumma, or rapidly growing malignant growth;
- over unhealthy skin, e.g., a varicose ulcer, an irritable patch of dermatitis;
- over a corn or callus, e.g., perforating or trophic ulcer;
- over a scar, e.g., Marjolin's ulcer.

- *Has the ulcer developed after sexual contact, e.g., chancre or chancroid?*

3. Duration

How long is the ulcer present there?

- Short in acute ulcer; long in chronic ulcer.
- Incubation period in venereal ulcer: 3 to 4 weeks in case of Hunterian chancre (syphilis) and 3 to 4 days in case of chancroid (soft sore).

4. Painful or painless

Is the ulcer painful?

- Inflammatory ulcers are painful.
- Tuberculous ulcers are mildly painful.
- Malignant ulcers are absolutely painless unless they, in late stage, infiltrate the adjacent structures supplied by the nerve endings.
- Syphilitic ulcers and trophic ulcers resulting from nerve diseases (tabes dorsalis, syringomyelia, transverse myelitis, peripheral neuritis) are painless.

5. Progress of the ulcer

Is it changing in size or surface?

6. Nature of discharge

- Serum, pus, or blood.

7. Past history

- Diabetes, tuberculosis, syphilis, history of exposure, or some neurological disease should be enquired.

Local examination

Inspection followed by palpation:

- Site

- Number, size and shape
- Margin
- Edge
- Floor
- Discharge
- Base
- Tenderness, friability or bleeding

Site

This is often typical, e.g., *rodent ulcer* on the upper part of the face; *tuberculous ulcer* on the neck; *trophic ulcer* over the weight-bearing area e.g., over the heel, over the sacrum or other bony point in a bed-ridden patient; *ischaemic ulcer* over the dorsum of the foot and toes; *varicose ulcer* on the medial side of the lower half of the leg.

The three common types of lower limb ulcer – venous, arterial and neuropathic ulcers have specific site predilection.

Number, shape and size

- Number: Single or multiple i.e., similar ulcers elsewhere in the body e.g. pressure sores.
- Shape: The ulcer may be round, oval, irregular, or serpiginous.
- Size: Measure the size in cm in two directions.

Margin

Margin of the ulcer is the border or 'transitional zone' of skin around the ulcer i.e., *it is the line demarcating the ulcer from the intact skin*. Three types of margins are encountered:

1. *Healing ulcer margin*: shows typical bluish line of growing epithelium which is squamous without cornification.

Margin of a healing ulcer shows three zones:

- (i) Outer – white
- (ii) Intermediate – blue
- (iii) Inner – red

2. *Spreading ulcer*:

- (i) Irregular in malignant ulcer, e.g. basal cell carcinoma
- (ii) Red, inflamed and irregular margin with inflamed surrounding tissue in infective ulcer.

3. *Chronic non-healing ulcer*: shows marked fibrosis with thick white skin margin without the blue line of growing epithelium.

Edge

Edge of the ulcer is the mode of union between the floor and the margin of the ulcer, and it has thickness in three dimensions. It can be inspected as well as palpated.

Edge is often characteristic of the underlying pathology:

1. *Slopping edge*: A healing non-specific ulcer, venous ulcer.
2. *Punched out edge*: Trophic ulcer. The tissue destruction is equal in all planes from skin to bone, so the ulcer becomes deep *with a vertical edge* as if the tissues have been punched out.
3. *Undermined edge*: In a tubercular ulcer, the tissue destruction is more in the subcutaneous plane than in the skin, so that the skin overhangs at the edge. This can be well demonstrated by passing a probe under the margin. This is termed as undermined edge.

Undermined edge is also noted in ulcer due to pressure necrosis particularly over the buttocks and in carbuncles.

4. *Raised edge*: Rodent ulcer. The ulcer edge is raised and rolled but not everted. This is a slow growing malignancy. There may be nodules on the raised edge of the ulcer – *beaded appearance*.
5. *Raised and everted edge*: Epithelioma. In a malignant ulcer, the malignant tissue grows very

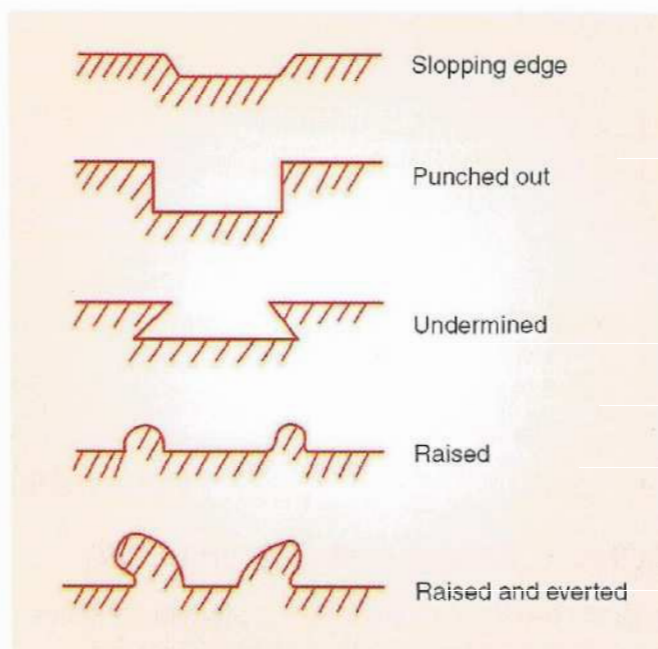


Fig. 1.1.: Edge of the ulcer.

fast and overhangs the skin margin. The ulcer itself is raised above the skin margin. The ulcer edge is raised and also overhangs the margin of the ulcer towards the surrounding skin – raised and everted margin.

Floor

This is exposed surface of the ulcer i.e., the part which can be seen within the edge of the ulcer. This may show:

- *Pink or red and granular granulation tissue.* No slough is present. There may be small amount of serous discharge. e.g., healing ulcer.
- *Pale, flat and smooth granulation tissue* which does not bleed easily on touch e.g., chronic or slowly non-healing ulcer.
- *Areas of unhealthy granulation tissue and areas of slough*, e.g., spreading or infective ulcer. Slough is the necrotic soft tissue which has not yet separated from the living tissue, e.g., infected bed sores.
- *Hypertrophic granulation tissue* where epithelialisation is not completed in time and thus shows hypertrophic granulation tissue. The exuberant granulation tissue rises above the skin surface. This is termed as sprouting flesh and is accompanied by excessive serosanguineous discharge. *Sprouting granulation tissue also delays wound healing*, e.g., a larger size ulcer of some duration.
- *Watery or apple jelly granulation tissue* e.g., tuberculous ulcer
- *Wash leather slough* e.g. a gummatous ulcer.
- *Malignant tissue.*
- *Relationship of the floor to the surface.*
 - (i) Floor below the surface level – non-malignant ulcer.
 - (ii) Floor above the surface level i.e., raised from the surface – malignant ulcer.

Discharge

This may be small or profuse. The type of discharge may be typical, e.g.

- Purulent discharge – indicates active infection.
- Greenish discharge suggests infection with *Pseudomonas pyocyanea*.
- Watery discharge – typical of tuberculosis.
- Sulphur granules – typical of actinomycotic ulcer.

Base

This signifies the area on which the ulcer rests. The base is to be palpated through the floor of the ulcer for:

- Mild induration due to fibrosis may be felt in any chronic ulcer.
- Marked induration is almost diagnostic of malignant ulcer.
Hunterian chancre also shows marked induration at the base.
- Mobility of the ulcer over the underlying structures – *reduced mobility implies* fixity to the underlying structures, muscle or bone. Malignant ulcer has reduced mobility. Varicose ulcer is also usually attached to the tibia.
If the ulcer is small, try to pinch it up and palpate the base between two fingers.

Friability

Too much friability is often diagnostic of malignancy.

Bleeding on touch

Suggestive of malignancy. May also be noted in presence of healthy granulation tissue.

Examination of the surrounding area

This may show important features:

- (i) If the ulcer is infected and spreading due to cellulitis, surrounding skin appears shiny, red and oedematous with tenderness.
- (ii) Dark pigmentation and eczema in a varicose ulcer.
- (iii) Multiple scars and puckering of the skin, sinuses surrounding the ulcer: suggestive of tubercular ulcer.
- (iv) Matted nodes surrounding the tubercular ulcer.
- (v) Melanotic halo in malignant melanoma.
- (vi) Satellite nodules within 5 cm proximal to malignant melanoma.
- (vii) A chronic ulcer within a large scar suggests Marjolin's ulcer.

Examination of the regional lymph nodes

The absence or presence and the consistence of the nodes may often indicate the type of the ulcer, e.g.

- Nodes are soft and tender in case of infective ulcer.
- Nodes are firm and matted and non-tender in Koch's ulcer.

- Nodes enlarged, hard and even fixed in case of carcinomatous ulcer.
- Nodes not enlarged in case of rodent ulcer unless secondarily infected.
- Inguinal nodes draining a syphilitic chancre of the penis are firm and 'shotty'.

When the regional nodes are palpable, then palpate the next higher groups of lymph nodes in a malignant ulcer.

General examination

1. Examination for debility and malnutrition – anaemia, diabetes, cardiac failure, etc. which delays ulcer healing.
2. *Examination for impairment of circulation:*
 - Absence of arterial pulsation – arteriosclerosis, Burger's disease, Raynaud's disease, diabetes – since *poor blood supply* may result in ischaemic ulcers and delayed healing.
 - Varicosity of veins in presence of varicose ulcer. Also test for deep vein thrombosis and calf muscle tenderness.
3. *Examination for neurological deficits*, e.g., diabetes, tabes dorsalis, spina bifida, peripheral neuritis, leprosy may give rise to ulceration *because of anaesthesia of the part* – neuropathic ulcer.
4. *Examine the joint movements active and passive*, close to the ulcer. Restriction of joint movements and presence of deformity indicates a painful inflammation and tendon/muscle involvement. Varicose ulcer may cause equinus deformity of foot.
5. *Examine abdomen* for splenomegaly in haemolytic anaemia – leg ulcers.

Pathological examination

1. Examination of blood
 - Hb% – Anaemia; RBC – sickle cell anaemia; WBC – infection.
 - ESR: Tuberculosis, malignancy, chronic infection.
 - Sugar: Diabetes.
 - W.R., Kahn and VDRL tests: Syphilis.
2. Examination of the discharge from the ulcer: Bacteriological examination of the discharge – smear test and culture and sensitivity test – are

often required to determine the nature of bacterial infection.

Spirochetes are found in the discharge or scraping of a primary chancre.

3. Biopsy
 - *Wedge biopsy:* Removing a wedge from the margin of the ulcer. The central part of the ulcer is not chosen as it contains mainly necrotic material.
 - *Excision biopsy:* The whole of the ulcer including the base may be excised and examined.
4. X-ray
 - Off the underlying bone.
 - Chest, in tubercular or malignant ulcer.

LIFE HISTORY OF AN ULCER

This consists of three stages:

1. *Stage of extension* i.e., spreading or sloughing.
2. *Stage of transition* i.e., preparation for healing.
3. *Stage of repair.*

Different characters of ulcers are evident in different stages.

Stage of extension

- Floor: Covered with exudates and sloughs. No granulation tissue.
- Discharge: Often purulent and even blood-stained.
- Edge: Sharply defined, thickened and inflamed.
- Surrounding area: Inflamed and oedematous.
- Base: Indurated and fixed.
- Slough and small amounts of discharge may dry to become a scab. A layer of dead tissue may become dehydrated and form a dark brown or black eschar such as after a burn or ischaemic necrosis.

Stage of transition

- Floor: Becomes cleaner with sloughs separating. Small, reddish areas of granulation tissue appear which link up ultimately to cover the whole surface.
- Discharge: Becomes serous.
- Base: Induration diminishing.

Stage of repair

Follows the transition stage and may either show signs of healing or characters of a callous ulcer.

(a) Signs of healing

- **Floor:** Contains smooth and even red granulation tissue covered by a single layer of epithelium. The granulation tissue is transformed into fibrous tissue which gradually contracts to form a scar.
- **Edge:** Becomes more shelving with the bluish epithelium gradually extending from the margin onto the floor of the ulcer to cover it up (at a rate of 1 mm per day).
- **Discharge:** It is merely serous, if the surface is kept at rest.
- **Surrounding skin:** It is soft, flexible and free from congestion.
- **Base:** It is free from fixity.

(b) Change to indolent or callous ulcer

This means that the ulcer refuses to heal by itself.

- **Floor:** Covered with unhealthy pale granulation tissue.
- **Edge:** Thickened, oedematous, indurated and often discoloured.
- **Surrounding area:** Oedematous and indurated.
- **Base:** Indurated.

In a malignant ulcer, the base is formed of malignant tissue. There is no formation of granulation tissue and skin ingrowth.

Typical features of some common ulcers**Epithelioma (Syn. Carcinomatous ulcer; Squamous cell carcinoma of skin)**

1. May occur anywhere but commonly found in lips, cheeks, tongue, breast, penis and anus.
2. Usually painless.
3. Irregular shape with rolled out and everted edges are typical.
4. Regional lymph nodes enlarged and hard.

Rodent ulcer (Syn. Basal cell carcinoma of skin)

1. Commonly found in the upper face above a line joining the angle of the mouth with the lobule of the ear.
2. Usually painless.
3. Usually circular in shape with raised and beaded edges are typical.
4. Minute venules in the edge of the ulcer are characteristic.
5. Regional nodes are not enlarged.

Tuberculous ulcer

1. Usually results from bursting of caseous lymph nodes, so commonly seen on the neck.
2. Usually painful.
3. The edge of the ulcer is thin and undermined and frequently bluish in colour.
4. There is pale granulation tissue at the floor with watery discharge. The base is mildly indurated.
5. Presence of matted lymph nodes adjacent to the ulcer.

Syphilitic ulcer (Syn. Gummatous ulcer)

1. Usually seen over the subcutaneous bone (e.g., tibia, ulna, sternum and skull).
2. Painless ulcer.
3. Most characteristic features are the punched out edge of the ulcer and the presence of wet wash leather (yellowish grey) slough in the floor.
4. Lymph nodes are seldom involved unless secondarily infected.

Hunterian chancre (Primary syphilitic sore)

1. Usually seen over the penis, lips and tongue.
2. Usually appears 3 weeks after the infection.
3. The ulcer is painless.
4. It is usually oval in shape with a slopping edge and exudes a blood-stained discharge. The base of the ulcer is characteristically indurated that feels like a button.
5. *T. pallidum* can be demonstrated in the serous discharge.
6. The lymph nodes in the groin are enlarged, and 'shotty', i.e., hard and small and painless with no tendency to soften or suppurate.
7. Extragenital chancres are frequently not indurated, and the involved lymph nodes are often considerably enlarged.

Soft chancres (Ducrey's)

1. Usually seen on the genitalia.
2. Usually appear within 3 days after infection.
3. Painful ulcers.
4. Present as multiple acute ulcers with oedematous edge and yellowish slough, discharging copious purulent secretion.
5. Involved lymph nodes show the picture of acute lymphadenitis with tendency towards suppuration.

Varicose ulcer

1. Typically situated on the medial side of the lower half of the leg.
2. It is *painless* callous ulcer.
3. The ulcer is vertically oval in shape with slopping or stepping edge. It never penetrates the deep fascia.
4. Presence of pigmentation or eczema in the skin around the ulcer.
5. Presence of varicose veins in the upper part of the leg or thigh clinches the diagnosis.
6. Ask the patient to stand and examine both long and short saphenous veins for varicosity. Also note for scattered and irregular varicosities, blow outs.
7. Varicose ulcer may cause equinus deformity of the foot.

Post-thrombotic ulcer

1. It follows deep vein thrombosis following parturition or operation.
2. A *painful ulcer*, situated on the lower leg. The ulcer always penetrates the deep fascia.
3. Do the test for deep vein thrombosis: calf muscle tenderness, Homan's sign and Mose's sign.

Trophic ulcer

A trophic ulcer is due to an impairment of nutrition to the tissues – either an impairment in blood supply, i.e., *ischaemia* or an absence of properly functioning nerve supply, i.e., *anaesthesia*. So there are two types:

1. Arterial ulcer
2. Neurogenic ulcer

Arterial ulcer (Ischaemic ulcer)

1. The ulcer is caused by inadequate blood supply to the tissues or skin, mostly due to peripheral arterial disease resulting in poor peripheral circulation. So, mostly found in presence of atherosclerosis (commonest), Burger's disease, Raynaud's disease, diabetes.
2. Commonly seen in old people.
3. Commonly occur in those parts of the limbs which are subjected to repeated pressure and trauma. Repeated trauma and superadded infection cause destruction of the poorly vascularised skin which fails to heal because of poor arterial blood supply.

4. Usually occur on the anterior and lateral aspects of the leg, on the toes, dorsum and sole of the foot, or the heel (these are the parts exposed to repeated pressure or trauma). May also occur on the finger tips and dorsum of the hand.
5. *Painful ulcer*. Moreover, a history of intermittent claudication with or without discoloration of the toes is mostly present and characteristic.
6. Ulcers may have punched out edges with slough in the floor. Very often the deep fascia is involved exposing the tendons in the floor of the ulcer (cf. venous ulcer).
7. Patches of dry gangrene may also be present along with the arterial ulcer.
8. Peripheral arterial pulses – diminished or absent. *Palpate all the related arteries of both sides to rule out arterial insufficiency.*
9. The ischaemic limb feels cold.
10. Elevation of the affected limb above the heart's level causes a marked pallor of the limb within 2–3 minutes and the patient will complain of pain in this position. Lowering of the limb below the horizontal leads to cyanotic congestion.
11. Angiography may be necessary to detect the arterial disease.

Neurogenic ulcer (neurotrophic)

1. The mechanism of formation of such an ulcer is repeated trauma or pressure *in an area which has absent sensation, i.e., anaesthesia due to impaired nerve supply.*
2. Such ulcers are found in presence of diabetic neuritis, peripheral nerve injury, spina bifida, leprosy, tabes dorsalis, syringomyelia. *Bedsore and perforating ulcers are examples.*
3. These ulcers are commonly seen on the buttock and on the back of the heel when the patient is non-ambulatory; on the heel and ball of the foot when the patient is ambulatory.
4. Ulcers may have punched out edges with slough in the floor. Very often the deep fascia is involved exposing the tendons and bones in the floor of the ulcer.
5. *Painless ulcer because of anaesthesia.*
6. The ulcer starts as a callosity under which suppuration takes place. The pus comes out from a small central hole and thus an ulcer is formed which gradually burrows silently through the

deep fascia, muscles and the tendons to bones and joints. *This is why this ulcer is also called perforating ulcer.* The resultant cavity becomes filled with offensive matter. Finally, the track becomes lined with unhealthy granulation tissue or skin thus healing becomes impossible.

7. Peripheral arterial pulsations are well palpable.
8. Neurological examination will reveal diminished or absent sensation. *Test the sensation of the skin surrounding the ulcer using a sharp pin.*

Diabetic ulcer

Three aetiological components are responsible for a diabetic ulcer:

1. Impairment of peripheral circulation leading to local ischaemia:
 - atherosclerosis – as diabetic patients are more prone to it at an earlier age;
 - microvascular disease.
2. Impairment of sensation due to peripheral neuropathy.
3. Poor resistance of the tissues to trauma and infection owing to the presence of sugar in the tissues.

Characteristics of ulcer are same as that of trophic ulcer.

Leprosy ulcer

Note the followings:

1. Hypopigmented anaesthetic patches over the limbs, back, face.
2. Features of leonine face.
3. Palpate: Posterior tibial, ulnar and greater auricular nerves for thickening.

Examine the deformity and the joint movements active and passive, close to the ulcer. Restriction of joint movements and presence of deformity indicates a painless inflammation and tendon/muscle involvement.

- Congenital sinus – Preauricular sinus.
- Acquired sinus – Pilonidal sinus.
- Osteomyelitic sinus discharging pus **with or without** bony spicules.

FISTULA

A fistula is an abnormal tract between two epithelial surfaces. *So, it has two openings.*

The communication may be between a hollow viscus and the skin, termed as *external fistula*; or between two hollow viscera, termed as *internal fistula*.

The tract is lined by either granulation tissue or epithelium.

- Examples of external fistula are anal fistula, umbilical fistula, parotid fistula, branchial fistula and thyroglossal fistula.
- Examples of internal fistula are vesicovaginal fistula, gastrocolic fistula, cholecystoduodenal fistula, tracheo-oesophageal fistula and recto/colovesical fistula.
- The communications may be between two vessels – arteriovenous fistula.

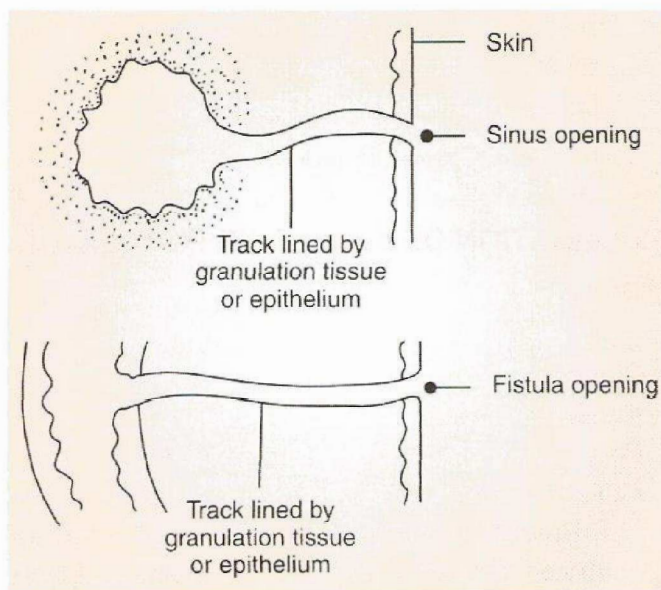


Fig. 1.2.

SINUSES AND FISTULAE

SINUS

A sinus is an abnormal *blind tract* leading from an epithelial surface into the surrounding tissue, *so it has one opening.* The sinus is lined by either unhealthy granulation tissue or epithelium. Examples of sinus:

Classification

Sinuses and fistulae may be congenital or acquired.

- *Congenital forms:* Preauricular sinus, branchial fistula, tracheo-oesophageal fistula, umbilical sinus, urachal sinus.

- *Acquired forms:* Often follow inadequate drainage of an abscess. Examples are perianal abscess bursting on the surface leading to external fistula, or opens both into the anal canal and onto the surface resulting in fistula-in-ano. Other examples are pilonidal sinus, osteomyelitic sinus, hydradenitis suppurativa.

Gastrojejunostomy and choledochoduodenostomy are caused by operation (iatrogenic).

Acquired arteriovenous fistulas are caused by trauma or by operation (iatrogenic arteriovenous fistula for renal dialysis).

Causes of persistence of a sinus or fistula

1. Presence of a foreign body or necrotic tissue (e.g., a suture, sequestrum, a faecolith or even a worm in the depth).
2. Inadequate or non-dependent drainage.
3. Prolonged discharge of irritating materials such as urine, faeces or bile causing persisting inflammation.
4. Associated with specific type of chronic inflammation, e.g., tuberculosis, actinomycosis, Crohn's disease, leprosy.
5. When the tract wall becomes epithelialised.
6. Presence of dense fibrosis around the tract prevents contraction and healing.
7. Unrelieved obstruction of the lumen of the viscus or tube distal to the fistula.
8. Presence of malignant disease.

EXAMINATION OF A SINUS OR FISTULA

History

Duration:

- Congenital
- Acquired

Past history of:

- Tuberculosis – tuberculous lymphadenitis followed by caseation of a lymph node, or abscess formation which had burst and discharge cheesy material.
- History of increasing swelling of the limb near a joint and then formation of an abscess which on bursting (or after operation) leaves a discharging sinus (suggestive of osteomyelitis).
- Previous anal abscess or ischiorectal abscess after inadequate drainage may lead to perianal fistula.

Local examination

1. *Number:* *Fistula-in-ano* may have more than one external opening particularly in presence of Crohn's disease affecting rectum and anal canal. 'Watering can' perineum with multiple openings can be seen in presence of urethral stricture. *Sinuses due to actinomycosis or madura mycosis are always multiple.*
2. *Positions:* Specific positions are often diagnostic of specific conditions, e.g., branchial fistula, preauricular sinus, pilonidal sinus, tuberculous sinus in the neck, etc.
3. *Visible opening of a sinus or fistula:* Sprouting granulation tissue at the opening of a sinus suggests a foreign body in the depth, e.g., a sequestrum, drainage tube or bullet. The opening of a tuberculous sinus is often wide resembling an ulcer with a thin, blue and undermined edge.
4. *Character of discharge:* It may be mucus, pus, blood, sulphur granules (actinomycosis), urine, faeces, bile, etc.
5. *Surrounding skin:* Presence of a scar may indicate chronic osteomyelitis or a previously healed tuberculous lesion. There may be presence of dermatitis, pigmentation.
6. *Palpation of a sinus or fistula for following:*
 - Tenderness
 - Thickening of the wall of a sinus – thickened in a chronic sinus.
 - Mobility of the sinus or fistula over the deeper structures – osteomyelitis sinuses are often fixed to the underlying bone.
 - Nature of discharge on pressure.
7. *Examination of the draining lymph nodes* – matted lymph nodes may be found in case of tuberculous sinus.
8. *Examination with a probe:* Probing should be done cautiously without using any force. The following points are noted:
 - The direction and depth of the sinus.
 - The presence of any foreign body or a movable sequestrum in the depth of the wound.
 - Whether the probe enters a hollow viscus or a bony cavity.
 - Whether any fresh discharge comes out on withdrawal of the probe.

General examination

1. Examination for debility and malnutrition, anaemia, diabetes – all may cause delayed healing.
2. Examination of particular system depending on the site and cause of sinus should be performed, e.g.
 - Chest in presence of chronic empyema.
 - Rectum and anal canal in presence of perianal fistula.
 - Urethra and lower urinary tract in presence of 'watering can' perineum.
 - Bone in presence of osteomyelitis.

Special investigations

1. Examination of the discharge – *physically* (e.g., cheesy material in tuberculosis), *chemically* (e.g., presence of urea suggests the diagnosis of a fistula of renal origin), *microscopically* (e.g., sulphur granules in actinomycosis), and *bacteriologically* (e.g., Gram stain, culture and sensitivity).
2. X-ray examination
 - Straight X-ray – to show a foreign body or a sequestrum and bony changes in osteomyelitis.

- *Sinogram or fistulogram* – injection of a radio-opaque dye (lipiodol or hypaque) into a sinus or fistula will determine the course and disposition of the sinus, its relation with the hollow cavities or viscus.
3. Often methylene blue dye is injected into the fistula or sinus to stain the tract – it helps in tracing the direction or disposition of the tract during operation.
 4. Biopsy from the edge of the sinus/fistula may reveal tubercular or malignant aetiology.
 5. Blood sugar to rule out diabetes.

Management of a sinus or fistula

- Correction of malnutrition, anaemia and diabetes.
- Control of infection with antibiotics – specific antibiotics may require Gram stain and culture sensitivity, Leishman stain.
- Adequate drainage and/or excision – may require radiological investigations. Methylene blue dye injection prior to operation helps in tracing the tracks.
- Sequestrectomy followed by saucerisation of the bone cavity for chronic discharging osteomyelitis.
- Removal of any foreign body.
- Adequate rest.

2

CHAPTER



Skin and Subcutaneous Tissue

PAPILLOMA OF THE SKIN

Papilloma is a hamartoma consisting of an overgrowth of all layers of skin and its appendages. It has a central core of connective tissue and blood vessels. A cutaneous papilloma may be derived from either the squamous or the basal cell layers.

Squamous cell papilloma

3 types are encountered:

1. *Congenital* (Syn.: Naevus verrucous) – appears either at birth or in early life and may be single or multiple. It usually presents as a brownish warty growth.
2. *Soft papilloma* – often seen on the eyelids of elderly people.
3. *Keratin horn* – a papilloma with excess keratin formation; common in elderly people (Fig. 2.1).

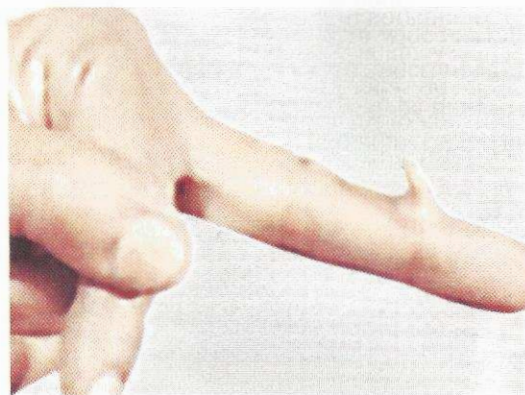


Fig. 2.1. Keratin horn dorsum of the middle finger.

6. Symptoms mostly give rise to cosmetic problem or can catch on clothing.
7. Treatment: Excision for cosmetic reasons.

Characteristic features

1. It can occur anywhere on the body.
2. It appears as well-defined swelling, usually pedunculated, ranging from a few millimetres to few centimetres.
3. Surface smooth or grooved or deeply fissured, and may be associated with pigmentation or keratosis.
4. Consistence – solid, usually firm, but may be soft due to excess fatty component.
5. Noninvasive and nonindurated unless inflamed and ulcerated.

Basal cell papilloma

(Syn.: Seborrhoeic keratosis, Senile warts)

It is due to overgrowth of the basal layers of the epidermis. It is a benign tumour.

Characteristic features

1. They usually appear as multiple lesions in the elderly people of both sexes.
2. Common in Caucasians and uncommon in Negroes and Indians.
3. They can occur in any area but most predominantly over the face and neck, and shoulders and trunk.

4. Usually the lesions look like an ovoid, verrucous, brown plaque stuck on the epidermis, and varies from few millimetres to few centimetres.
5. Surface is rough and hyperkeratotic and has a waxy look and feel.
6. Slowly growing lesion and may fall or be rubbed off.
7. May become infected and ulcerated. Malignant transformation has been recorded. It is also considered to be a marker of visceral malignancy. When deeply pigmented, it is mistaken for a malignant melanoma.
8. Symptoms: Mostly give rise to cosmetic problem or can catch on clothing.
9. *Treatment:* Removal for cosmetic reasons.
 - The lesion can be scraped with a sharp curette which leaves a flat surface which becomes covered with normal epithelium in one week.
 - Cryosurgery is also useful.

WARTS

A wart is a rough excrescence on the skin due to infection with the papilloma virus of the papovavirus group. A number of different viruses are involved. It is transmissible through direct or intimate contact with inoculation through a minor abrasion.

Characteristic features

1. They occur at all ages but common in children and young adults.
2. They usually tend to occur on sites of trauma, such as the beard area, hands, genital region and feet.
3. All warts first appear as small, smooth nodules, later they may coalesce to form rough excrescence.
4. They may be pigmented, keratinised or deeply pitted with a frond-like surface.
5. Warts can cause pain and irritation.
6. Plantar warts occur on the sole. They are inverted into the skin and, due to the pressure of walking are painful and tender.
7. Warts can cause cosmetic problems when on the hands. Multiple warts in the fingers may interfere with the fine movements.
8. Venereal warts (papilloma acuminata) are seen in the coronal sulcus or the perineum. They are multiple and moist with offensive discharge.

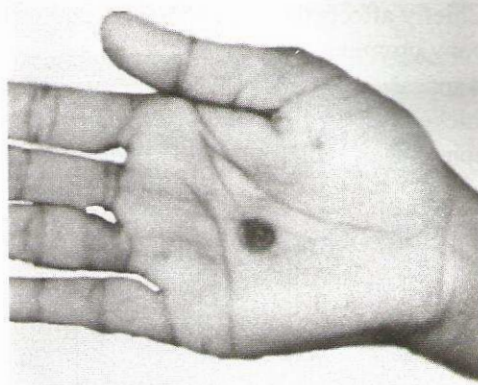


Fig. 2.2. Palmar wart.

9. Two-thirds of warts may disappear spontaneously.
10. Treatment:
 - Curettage and diathermy
 - Repeated application of 20% podophyllin in liquid paraffin.
 - Cryosurgery.

CALLOSITIES

1. A callosity is a localised thickening of the skin due to excessive keratinisation.
2. It occurs as a protective measure at the sites of friction.
3. It appears as superficial, circumscribed, yellow white patches of hyperkeratotic material and is continuous around its margins with the normal skin. *It is usually painless.*
4. Commonly seen in the feet, especially the infra-medial aspect of the great toe, and the heel. It may also occur over the dorsal aspect in other limb deformities particularly neuropathic. Also found in gardeners' hands.
5. Histologically there is thickening of the stratum corneum and granulosum with or without atrophy of the rete pegs.
6. Treatment is usually not required.

CORN

1. A corn is a localised hyperkeratinisation of the skin with a hard central core.
2. It occurs due to undue pressure as seen with ill-fitting or tight shoes.

3. Chiefly affects the toes and feet, usually over a bony prominence.
4. It appears as a circumscribed cone-shaped horny thickening with the base on the surface and apex pointing inwards. *The lesion is usually painful and tender.*
5. Treatment:
 - Preventive: Use of ill-fitting or tight shoes should be stopped. Soft shoes with soft pads at the pressure points to be used.
 - Local application of salicylic acid in collodion on successive nights may be useful.
 - If conservative measures fail, excision of the corn with particular care to remove the apex of the corn is required.

One should be careful in treating corn in patients with diabetes or with a poor peripheral circulation when a secondary infection may precipitate gangrene. Peripheral pulses examination and blood sugar estimation is mandatory before treating corns in elderly people.

DERMOID CYST

Synonym: Epidermal cyst

Clinically four varieties of dermoid are recognised:

1. Sequestration dermoid
2. Implantation dermoid
3. Tubulodermoid
4. Teratodermoid

SEQUESTRATION DERMOID

It is a true variety of congenital cyst arising from the primitive ectodermal cells being buried at the line of embryonic fusion. The cyst is lined by squamous epithelium and contains paste-like desquamated material.

Origin

During embryonic fusion a few ectodermal cells are sequestered into the deeper layer, i.e., into the surrounding mesoderm. Later on these cells proliferate and liquefy to form cystic swelling. During enlargement of the cyst the surrounding mesoderm can be pressed or indented giving rise to bony indentation (Fig. 2.3).

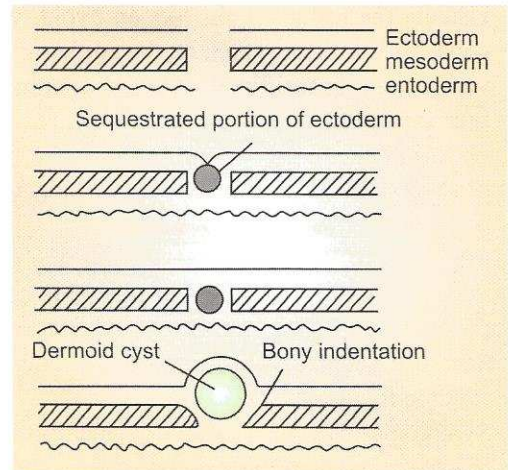


Fig. 2.3. Mode of formation of sequestration dermoid.

Sites

The planes of fusion are the usual sites of dermoid cysts, e.g.

- at the midline of the body – especially in the neck, at the root of the nose (Fig. 2.4);
- on the scalp – pteryon, asteryon (post-auricular), etc. (Fig. 2.8);
- at the inner or outer angles of the eye (angular dermoids) (Figs. 2.5, 2.6, 2.7).

Pathology

Lining of the cyst: Squamous epithelium with hair, hair follicles, sweat and sebaceous glands (cf. implantation dermoid).

Contents: Toothpaste-like desquamated material with or without hairs. The paste is the mixture of sebum, sweat and desquamated epithelial cell debris.

Diagnostic criteria

1. Painless lump at the common sites mentioned above.
2. Shape and size – ovoid or spherical swelling varying from 1 to 2 cm in diameter.
3. Cystic swellings – fluctuation positive.
4. Surface is smooth.
5. Skin can be lifted up (cf. sebaceous cyst).
6. Absence of punctum (cf. sebaceous cyst).
7. Not compressible (cf. meningocele).
8. The dermoid cyst itself may indent on pressure over the surface (cf. sebaceous cyst) (Fig. 2.5A&B).

DERMOID CYST

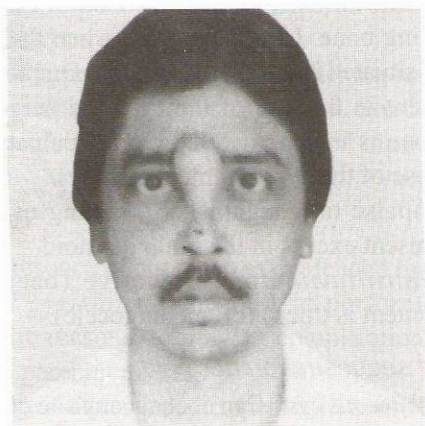


Fig. 2.4. Dermoid at the root of the nose.

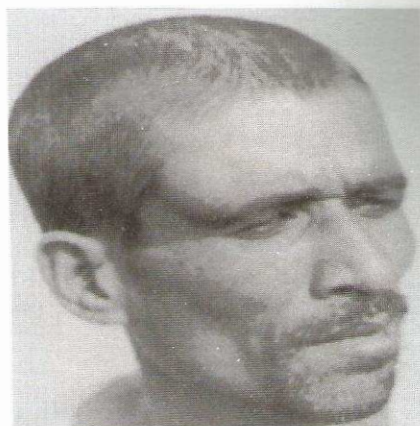


Fig. 2.6. Angular dermoid.



Fig. 2.5 A



Fig. 2.7. Angular dermoid.



Fig. 2.5 A & B. Angular dermoid. Note indentation over the swelling after finger tip pressure (cf. sebaceous cyst).



Fig. 2.8. Post-auricular dermoid.

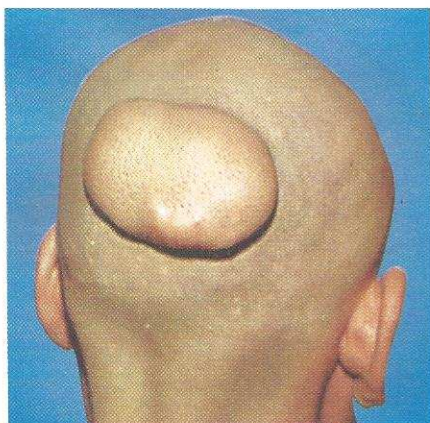
DERMOID CYST (Contd.)

Fig. 2.9. Recurrent dermoid over the occipital region. Note the scar from previous operation.

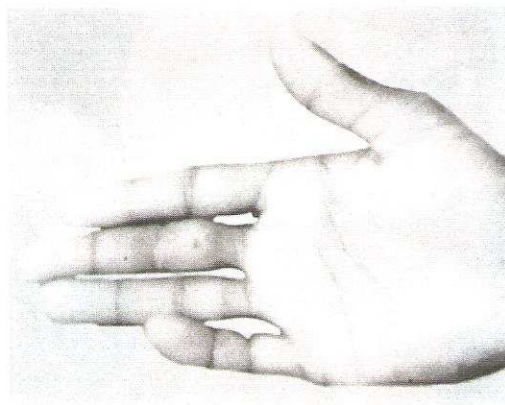


Fig. 2.10. Implanted dermoid right middle finger. Note puncture scar at the top of lump – 3 months duration.



Fig. 2.11. Implanted dermoid over right palm. No puncture scar noted over the swelling.

9. *Bony indentation*: Palpate the margin of the cyst which may reveal an indentation in the underlying bone. Usually present when the dermoid is situated in relation to a bone. Due to enlargement in the size of the cyst, the neighbouring bone is indented which can be palpated at the base of the cyst (cf. meningocele).
10. Impulse on coughing or straining usually not present except in the scalp dermoid.
11. *Transillumination*: Negative (because the content is thick) (cf. meningocele).

D/D of sequestration dermoid

1. *Sebaceous cyst*: Can occur at any site of the skin. *Punctum* is present. Dermoid cyst occurs along the lines of embryonic fusion. *Punctum* is absent.
2. *Meningocele*: Brilliantly transilluminable.
3. *Lipoma*: Slipping sign at the edge is present.

Peculiarities of scalp dermoid (Fig. 2.9)

1. Dermoid cyst may lie fully outside the cranial bones.
2. It may lie completely inside the cranial bones – between the dura and the cranial bones.
3. It may be partly intracranial and partly extracranial with a connection between them (Hourglass dermoid).
4. It may lie extracranially, but may be attached to the underlying dura by a stalk or pedicle passing through a defect in the bone.

The last two varieties only may show expansile impulse on straining or coughing when they have to be differentiated from meningocele by transillumination test which will be positive in meningocele but negative in dermoid.

Complications

1. Infection
2. Suppuration
3. Bursting
4. Ulceration
5. Cosmetically it looks ugly. When found in children, it causes parental distress.
6. Pressure effects on adjacent structures particularly with intracranial extension.

Investigation

Scalp dermoids should be X-rayed, particularly those showing impulse on straining or coughing. If there

is an appreciable gap, the operation can be delayed till adolescence to give the bony defect an opportunity for spontaneous closure.

In the presence of persistent gap after adolescence, an intracranial extension is suspected. So, CT scan is performed to assess the extent of intracranial extension.

Treatment

- In the absence of bony gap as evidenced by X-ray, *simple excision* of the cyst is performed for cosmetic reasons, to prevent complications or to relieve parental distress.
- If the age is advanced, the operation should be very carefully done, and before excising the cyst, it should be differentiated from meningocele.
- In the presence of intracranial extension as detected by CT scan, both extracranial and intracranial parts are excised by raising an osteoplastic flap or by craniotomy.

IMPLANTATION DERMOID

(Post-traumatic dermoid)

It is an acquired cyst arising from the indriven epithelium beneath the skin following a trauma, *particularly a puncture injury*. The patient may not remember the injury.

Sites

Usually found in the areas subject to repeated trauma, e.g., in the pulps or tips of the fingers, palm and sole. So, it is common in the tailors or women with the habit of sewing (prone to needle prick); also noted in gardeners (prone to thorn prick) (Figs. 2.10, 2.11).

Pathology

Lining of the cyst: Squamous epithelium but hair, hair follicles, sweat and sebaceous glands are absent (cf. sequestration dermoid). So, it is a true cyst.

Contents: White greasy material which is desquamated epithelial cells with mucoid degeneration.

Diagnostic criteria

1. Spherical smooth swelling, usually 0.2 cm to 1 cm in diameter, in the areas subjected to repeated trauma.
2. The cyst feels tense and hard, sometimes stony hard. It may be painful and tender.

3. The overlying skin is often scarred.
4. The history of an old injury with or without a scar overlying the cyst is the most significant diagnostic feature.

Complications

1. The cyst on the finger or palm may interfere with the grip and touch.
2. The cyst on the sole may cause limping.
3. Infection.
4. Cosmetically looks ugly.

Treatment

Complete excision: To remove inconvenience at work, to remove pain or tenderness or for cosmetic reasons.

TUBULODERMOID

This is a cystic swelling arising from the non-obiterated portion of a congenital ectodermal duct or tube. The swelling is due to the distention caused by the accumulation of the secretion of the lining epithelium of the patent segment of the duct.

Examples

1. Thyroglossal cyst – commonest example.
2. Post-anal dermoid – remnant of neuroenteric canal.
3. Ependymal cyst in the brain – remnant of neuro-ectoderm.

TERATOMATOUS DERMOID

This is a cystic swelling arising from the totipotent cells with ectodermal preponderance.

Common sites

1. Ovary – ovarian cyst.
2. Testis – teratoma.
3. Mediastinum
4. Retroperitoneal
5. Pre-sacral area.

} Usually appear as solid swelling rather than cystic

Contents

Usually contains derivatives of mesodermal elements such as cartilage, bone, tooth, hair, and also cheesy material.

SEBACEOUS CYST

This is a retention cyst due to accumulation of sebum resulting from obstruction to the duct of the sebaceous gland.

Anatomically, sebaceous glands are situated in the dermis and their ducts open either on to the skin surface directly or open into a hair follicle.

Sites

It can occur anywhere in the body wherever sebaceous glands exist. *Common sites* are the scalp, face, scrotum, vulva.

It never occurs in the palm and sole, because there are no sebaceous glands in these regions.

Pathology

Lining of the cyst: Squamous epithelium as it is considered pathologically epidermoid cyst.

Contents: Yellowish white poultice-like material with an unpleasant smell. Such material is a mixture of sebum, fat, desquamated epithelial cell debris.

Rarely, on histological section, an organism called *Demodex folliculorum* may be found, because these organisms are harboured in the wall of the sebaceous gland.

Diagnostic criteria

1. Swelling in a common site as mentioned above.
2. Shape – globular or hemispherical.
3. Size – varies from a few millimetres to 4–5 cm.
4. Presence of bluish black spot, called *punctum* indicating the site of blockage at the opening of the duct, may be noticed (Figs. 2.12 and 2.13).
The punctum is adherent to the cyst. On squeezing the cyst putty-like or cheesy sebaceous material with an unpleasant smell may come out through the punctum.
5. Skin cannot be lifted up (cf. lipoma, dermoid cyst).
6. Consistence – cystic or tense cystic, hence *fluctuation may be positive*.
7. Surface is smooth.
8. Not compressible.
9. Because of its putty-like consistency, *the swelling can often be indented by a finger tip* (cf. dermoid cyst).
10. *Transillumination* – negative, because the content is thick.

Differential diagnosis

1. Dermoid cyst.
2. Lipoma.
3. Meningocele.

Features of the scalp sebaceous cyst (cf. Scalp dermoid) (Fig. 2.14)

- (a) Swelling is free from underlying structure. Sebaceous cyst moves with the movement of the scalp.
- (b) No punctum is usually visible.
- (c) Usually loss of hair is noted over the swelling.
- (d) Skin is attached to the swelling (cf. dermoid).
- (e) No impulse on straining or coughing (cf. meningocele).
- (f) No bony indentation can be felt (cf. dermoid).
- (g) But the swelling itself *can be indented* by a fingertip pressure – this is diagnostic.

Features of scrotal sebaceous cysts (Figs. 2.15 & 2.16)

Scrotal sebaceous cysts are usually multiple and when well formed they feel solid. No punctum is usually visible.

Complications

1. Cosmetically looks ugly.
2. Infection and suppuration (Fig. 2.17): The cyst is commonly infected. The surface skin becomes red and inflamed. The cyst becomes tense and tender and on squeezing the cyst, pus may be expressed through the punctum. Repeated infection is a common phenomenon which makes the operation difficult. In the presence of infected sebaceous cyst, diabetes should be excluded in adults.
3. Ulceration.
4. Calcification (Fig. 2.15).
5. *Cock's peculiar tumour*: It is nothing but suppurating and ulcerating sebaceous cyst and looks like squamous cell carcinoma. Infection → suppuration → bursting → escape of sebaceous material and purulent material on the surface → chronic irritation leading to granulating ulcer or a granuloma looking like a tumour. The granuloma consists of histiocytes and foreign body giant cells (cf. Epithelioma).

SEBACEOUS CYST



Fig. 2.12. Sebaceous cyst at the back of the trunk. Note the blue punctum which is diagnostic of such cyst.



Fig. 2.15. Scrotal sebaceous cyst with calcification.

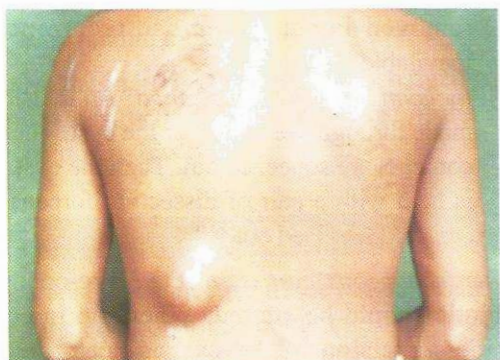


Fig. 2.13. Large sebaceous cyst on the back. Note the punctum at 12 o'clock position.



Fig. 2.16. Multiple sebaceous cysts involving the antero-inferior part of the scrotum – treated by partial excision of skin containing the cysts followed by primary suture of skin margins.

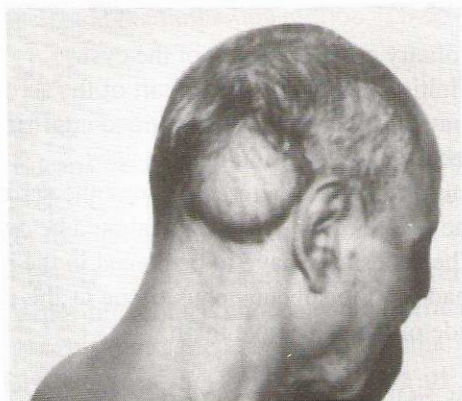


Fig. 2.14. A large sebaceous cyst of the scalp over right occipital region behind the right ear. Note the loss of hair over the surface of the swelling.



Fig. 2.17. Infected sebaceous cyst. Treated conservatively with antibiotic – the infection subsided and the swelling reduced much in size. Waiting for excision during quiescent phase. In the presence of infection, cyst removal is difficult and incomplete.



Fig. 2.18A. Sebaceous horn arising from the front of the neck. (Courtesy: Dr. R.L. Modak, MS)



Fig. 2.18B. Sebaceous horn on the dorsum of the forearm – just proximal to wrist.



Fig. 2.19. Sebaceous adenoma arising in the skin of upper back. Requires histological confirmation after excision.

6. *Sebaceous horn* (Figs. 2.18A & B): Impassated sebaceous materials may leak through the punctum and accumulate on the surface skin in successive layers. They get dried up and ultimately give rise to a horny projection.
7. Rarely malignancy occurs when it will be usually of basal cell carcinoma.

Treatment

Excision of the cyst either by dissection, or by incision and avulsion of the cyst wall.

- (a) *Dissection method*: An elliptical incision is made on the skin centering the punctum. Then the cyst with overlying skin is gradually dissected from the surrounding skin and subcutaneous tissue till the entire cyst can be removed intact, otherwise recurrence is inevitable.
- (b) *Incision and avulsion method*: An incision is made through the skin over the cyst. The cyst contents are squeezed out. Then the cyst wall is held with a pair of dissecting forceps and carefully avulsed out.

When sebaceous cyst is infected, pre-operative antibiotic should be started. When the infection subsides, excision of the cyst is performed. If infection does not subside, the cyst should be incised and the pus and semiliquid foetid material are expelled. This is followed by dissection or avulsion of the cyst wall.

Treatment of scrotal sebaceous cyst:

1. Solitary cyst – excision of the cyst.
2. Multiple cysts affecting a part of the scrotum – that part of the scrotal skin containing the sebaceous cysts should be excised.
3. Multiple cysts scattered all over the scrotum – the whole of the scrotal skin should be excised. The testes may have to be placed in the pocket made in the subcutaneous tissue at the medial side of the respective thigh.

SEBACEOUS ADENOMA (Fig. 2.19)

It is a benign tumour arising from the sebaceous gland. Histologically, simple hyperplasia of the cells occurs (epiloia). Clinically, it is a solid swelling, may be single or multiple, occurring either in the scalp

commonly in the frontal region, or in the regions of the nose and cheek. When occurring in the scalp, this may be associated with some lesion in the brain, even epilepsy (tuberculous sclerosis of the brain). When occurring on the nose, it is called rhinophyma.

Note: Sebaceous gland is a holocrine variety of exocrine gland producing secretions by fatty degeneration of its central cells.

Exocrine glands – three varieties:

1. Holocrine: Where the whole of the cell disintegrates and dies to liberate its secretion, e.g., sebaceous gland.
2. Apocrine: Where only the luminal part of the cell disintegrates leaving the nucleus and the basal portion from which the cell regenerates, e.g., mammary gland.
3. Merocrine: The secretion is discharged without any destruction of the cell. Most of the glands belong to this type.

LIPOMA

It is a benign, connective tissue tumour arising from the fat cells. It is the commonest of all benign tumours. It can occur in any situation where there is fat, hence known as universal tumour.

Common sites

The subcutaneous tissues over the trunk, the nape of the neck, and the limbs.

Varieties

In relation to possessing a capsule:

1. Encapsulated variety: Commonest. Lipoma is covered by a capsule and may occur at any site of the body. This is a true lipoma.
2. Diffuse variety: Rare. It is characterised by deposition of fat without any peripheral capsule, hence often called as 'pseudolipoma'. It does not possess the typical features of lipoma. Commonly found in people taking excessive alcohol, at the region of the neck extending to the face. The treatment here is nothing but to excise the excessive fat provided the patient needs that for cosmetic reasons.

Classification of lipoma

Anatomical classification

Depends on the anatomical sites:

1. Skin – Subcutaneous.
2. Aponeurosis or fascia – Sub-aponeurotic or sub-fascial.
3. Muscle – Intermuscular.
4. Periosteum – Subperiosteal.
5. Synovial membrane – Subsynovial.
6. Joints – Intra-articular.
7. Mucous membrane – Submucous.
8. Serous – Subserous (retroperitoneal).
9. Meninges – Extradural or subdural.
10. Gland – Intraglandular.

Histological classification

Depends on the presence of other tissues in addition to adipose tissue.

1. *Fibrolipoma* – mixture of fibrous tissue and adipose tissue.
2. *Naevolipoma* – mixture of haemangiomatous and adipose tissues.

Features of naevolipoma:

- Bluish discoloration of the overlying skin.
- Blanching.
- Compressibility.

3. *Neurolipoma* – mixture of nerve tissue and adipose tissue. It is often painful.

When they are multiple with pain and tenderness, it is called neurolipomatosis.

When neurolipomatosis is associated with the painful, tender, diffuse or nodular deposits of fat, it is called *Dercum's disease* or *adiposis dolorosa*. Commonly found in women, particularly affecting the trunk, buttock and thighs. Usually, no capsules are formed around these fatty deposits and so, these are called false lipomas.

Clinical classification

- | | | | |
|-------------|----------|-----------------|---------|
| 1. Solitary | } Number | 1. Sessile | } Shape |
| 2. Multiple | | 2. Pedunculated | |

Pathology

On cut section lipoma appears as uniform yellow-coloured fatty tissue traversed by white fibrous septa of varying thickness. Occasionally the cut surface may show patches of calcification and areas of myxomatous degeneration.

SUBCUTANEOUS LIPOMA

Diagnostic criteria

1. Painless, *soft, solid swelling* occurring at the common site.
2. *Surface usually is lobulated.* The lobules can be seen and felt on the surface and at the edge of the lump (Fig. 2.27B).
3. Edge is well-defined and may have a series of irregular curves corresponding each lobule.
4. Overlying skin is free (cf. sebaceous cyst).
5. *Slipping sign* – the edge of the swelling slips under the examining finger.
6. Freely mobile on both axes.
7. Cutaneous dimples appear on the surface when the swelling is pushed away (due to fibrous strand which traverses the lipoma and is attached to the overlying skin).
8. *Fluctuation – negative*; but due to its soft consistence there may be a false sensation of fluctuation known as pseudo-fluctuation.
9. *Transillumination – negative.*
10. Usually painless. When painful and tender, diagnosis of neurolipoma should be considered.

Complications

1. Cosmetically looks ugly.
2. Necrosis due to repeated trauma – the lump becomes hard and painful. The overlying skin may become tethered or fixed to the underlying area of fat necrosis.
3. Calcification.
4. Haemorrhage.
5. Infection.
6. Lipomatosis: Multiple contiguous lipomas may cause huge enlargement and deformity. Lipomatosis may occur in the limbs, neck, trunk and buttocks. Individual swellings within are identical to solitary lipomas.
7. Rarely malignancy – liposarcoma.
8. Myxomatous degeneration.
9. Ulceration.

Common sites where lipoma undergoes malignancy

1. Retroperitoneal lipoma – probably arises de novo.
2. Lipoma in the thigh, particularly the intermuscular type.
3. Subcutaneous lipoma of shoulder region.

Treatment

Excision, particularly if the lipoma is large, unsightly, or troublesome.

Characteristics of other varieties of lipoma

Subfascial lipoma: Lipoma may occur beneath the palmar or plantar fascia and is often mistaken as tuberculous tenosynovitis. In the palm it can cause median nerve compression.

Such lipomas may also occur in the areolar tissue beneath the epicranial aponeurosis of the scalp. This can erode the underlying bone and hence can be confused with a dermoid cyst.

Treatment: Early excision.

Intermuscular lipoma: Here lipoma occurs between the intermuscular fibres. Such lipoma becomes firmer on feel when the adjacent muscle contracts. It can cause mechanical interference with movement of the involved muscle. Intermuscular lipoma is mostly common in the thigh or around the shoulder. Fibrosarcoma and liposarcoma are also common in such sites and are difficult to differentiate from this condition clinically.

Treatment: Early excision to exclude fibrosarcoma/liposarcoma or to prevent mechanical interference.

Subsynovial lipoma: Such lipoma occurs in the fatty pad deep to the synovial membrane. Commonly seen in the knee joint when it should be differentiated from semimembranosus bursa or Baker's cyst.

Subserous lipoma: Rare and found beneath the pleura or peritoneum. Retropleural lipoma may present as a benign thoracic tumour. Retroperitoneal lipoma is more common in children than adults, often presents as an abdominal mass and is confused with hydronephrosis, pancreatic cyst, or teratomatous cyst. It may attain a very large size with heterogeneous surface.

Occasionally one may find a lipomatous mass at the fundus of the sac of a femoral hernia or epigastric hernia. This is nothing but condensation of extra-peritoneal fat rather than a true lipoma (see Epigastric hernia).

Submucous lipoma: It may occur in the respiratory tract when it may cause respiratory obstruction, or in the alimentary tract when it may lead to intussusception. It may also occur in the tongue when it can lead to macroglossia.

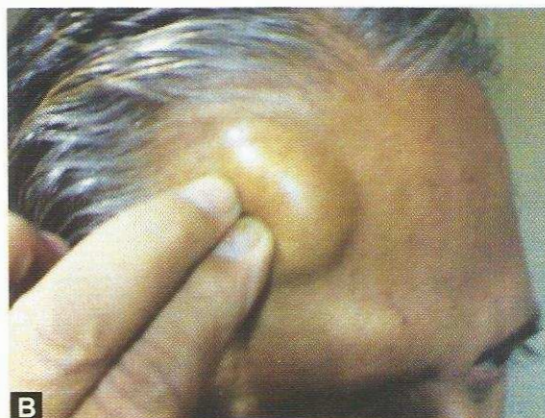
LIPOMA

Fig. 2.20A & B. Lipoma forehead (A). Margin slipped under the fingers (B).



Fig. 2.21A



Fig. 2.21A, B & C. Lump appeared to be angular dermoid on inspection (A). Margins of the lump slipped under the finger (B & C). But no indentation noted on finger tip pressure. So, lipoma. D/D: Angular dermoid.

LIPOMA (Contd.)



Fig. 2.22A. Lipoma. D/D: Angular dermoid.



Fig. 2.22B. Same patient. On inspection – the swelling appeared to be angular dermoid. But on palpation – a soft swelling; the swelling was not smooth; the edge of the swelling slipped under the finger. So, the diagnosis was lipoma. Diagnosis confirmed after operation. Note: Lipoma can occur anywhere in the body.



Fig. 2.23. Lipoma. Lobulated appearance and feel. The margin slipped under the finger. 10-year duration, gradually increasing in size. The overlying scar mark was due to application of Ayurvedic poultice.

D/D: 1. Angular dermoid – but the swelling not present since birth; no bony indentation with this size of swelling. 2. Sebaceous cyst – but no punctum; no surface indentation on finger tip pressure.

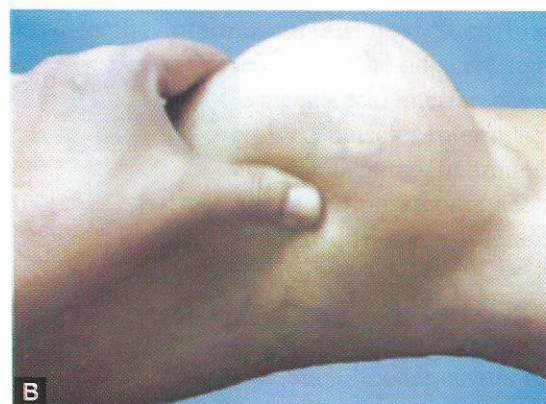


Fig. 2.24A, B & C. Lipoma on the medial side of the left thigh above the knee for nearly 10 years gradually increasing in size. Soft, solid, lobulated and freely mobile lump both over the relaxed and contracted muscles of the thigh as the lump was over the deep fascia of thigh. Margins slipped under the finger. (cf. Fig. 2.31 Liposarcoma)

LIPOMA (Contd.)

Fig. 2.25. Lipoma over the left buttock. Soft, solid, lobulated and freely mobile lump. Margins slipped under the finger. D/D: Liposarcoma. (Courtesy: Dr. Sudeb Saha, MS, FAMS)



Fig. 2.26. Lipoma left axilla – soft, solid, lobulated and freely mobile lump. D/D: Accessory breast. (Courtesy: Dr. Sudeb Saha, MS, FAMS)



Fig. 2.28. Lipoma right inguinal region. On inspection – the lump appeared to be inguinal hernia. But H/O persistent swelling was present. On palpation – a soft swelling; surface was not smooth; the edge of the lump slipped under the finger; there was no cough impulse and the lump was not reducible. So, the diagnosis was lipoma which was confirmed after operation.

Note: Lipoma can occur anywhere in the body.

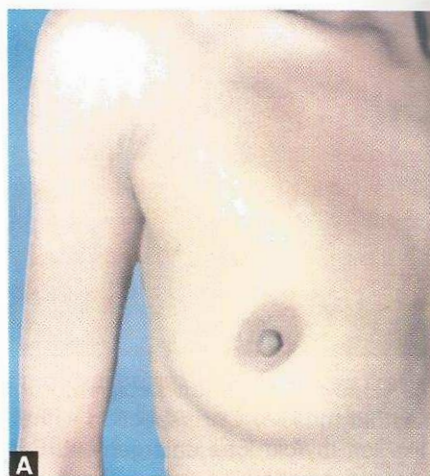


Fig. 2.27A. A soft swelling with indistinct surface lobulation noted over the right infraclavicular region. Margins were ill-defined and slipped under the finger.

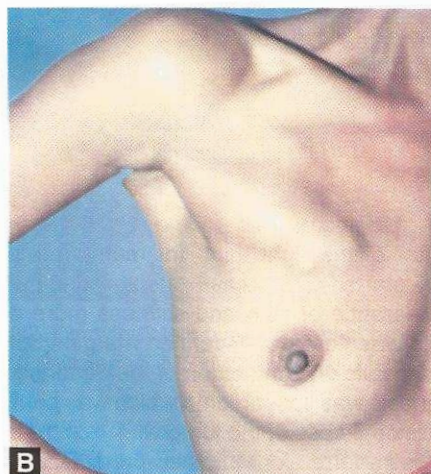


Fig. 2.27B. Same patient. Patient was asked to press on the hip – on contraction of the pectoralis major muscle, the swelling became more prominent. Note the irregular wavy margins and prominence of surface lobulation. The margins slipped under the finger – so the diagnosis was lipoma.

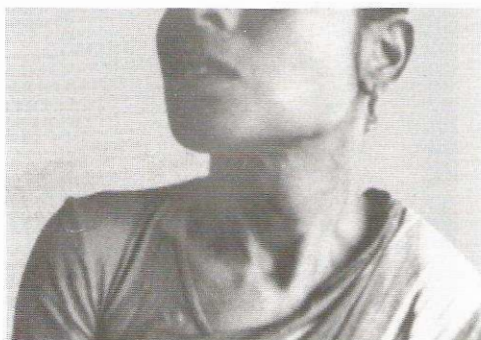
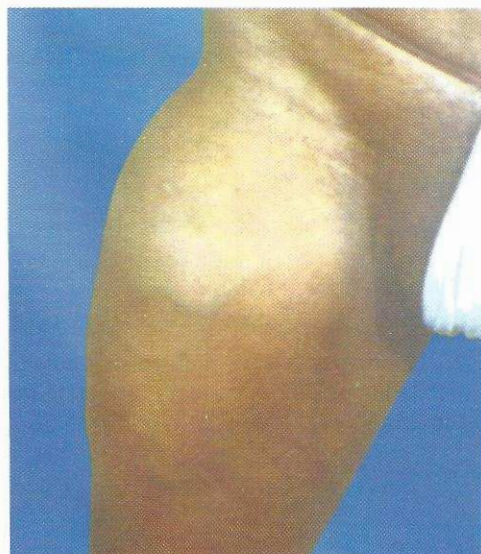
LIPOMA (Contd.)

Fig. 2.29. Lipoma in the left neck anteromedial to sternomastoid muscle. On inspection – the swelling looked like left thyroid lobe enlargement. But upward movement on swallowing was absent. Moreover, palpation revealed soft swelling with finely lobulated surface and irregular margin. On contraction of the sternomastoid the swelling became prominent and the margin slipped under the finger. So, diagnosis was lipoma and confirmed after operation. Note: Lipoma can occur anywhere in the body.



Fig. 2.30. Lipoma in the palm. Firm swelling in the proximal palm. No clinical evidence of median nerve compression (cf. carpal tunnel syndrome). D/D: Tuberculous tenosynovitis.

Fig. 2.31. Lipoma or liposarcoma (?). H/O soft swelling in the upper part of the right thigh for many years. The swelling suddenly started to increase in size during last six months with appearance of pain and stiffness more during walking. The lump was firm with ill-defined margins but became hard and less mobile on contraction of the quadriceps muscle. So, the diagnosis was liposarcoma which was confirmed after the total excision biopsy of the lump with 1 cm margin clearance. Patient was referred for radiotherapy. (D/D: Fibrosarcoma)



Extradural lipoma: This is a type of spinal tumour. Intracranial lipoma does not occur as there is no fat in the extradural tissue inside the skull.

Intraglandular lipoma: There are three glands in which a lipoma may occur:

- the breast,
- the pancreas, and
- beneath the renal capsule.

HAMARTOMA

It is a tumour-like developmental malformation of various tissues of which the part or the organ is made

of. It is characterised by their improper distribution with the prominence of one particular tissue.

The word 'hamartoma' means "missing the mark in spear throwing".

Characteristics of hamartomas

1. Developmental anomaly.
2. Present and often visible at birth.
3. During childhood they grow at the same rate as the surrounding tissues.
4. Enlargement continues until physiological growth ceases (i.e., on reaching adulthood).
5. Some may regress spontaneously when the tumour growth ceases, e.g., strawberry angioma.

6. May be multiple, e.g., neurofibroma.
7. Essentially a benign condition which very rarely undergoes malignant change.
8. *Different types of hamartomas* are found involving the blood vessels (haemangiomas), the peripheral nerves (neurofibroma and its variants), the intestinal tract (congenital polypi), the skin (pigmented naevus or mole), the lymphatics (lymphangioma), and in many other organs. Exostoses, lipoma, thyroglossal, branchial and dermoid cysts are also considered by some as hamartomas.

HAEMANGIOMA

A haemangioma is regarded as developmental malformations of blood vessels rather than a true tumour. It is an example of hamartoma.

It commonly occurs in the skin and subcutaneous tissues, but can occur in any part of the body, e.g., lip, tongue, mouth, internal organs like liver, lung, brain, intestine, etc.

Types

Three types are encountered according to the nature of blood vessels involved:

1. Capillary.
2. Venous or cavernous.
3. Arterial or plexiform.

A pure form is rarely seen and an admixture of different types is common. Clinically, capillary and cavernous varieties are commonly recognised.

CAPILLARY HAEMANGIOMA

Includes the following varieties:

- (i) Salmon pink patch
- (ii) Portwine stain
- (iii) Strawberry haemangioma
- (iv) Spider naevus.

I. Salmon pink patch

Characteristic features

1. Present since birth.
2. Commonly present as pink patch over the forehead in the midline, and over the occiput.
3. The appearance resembles the 'stork-bites'.
4. Usually disappears by the age of one year.
5. Hence, no treatment is required.

II. Portwine stain (naevus flammeus)

(Fig. 2.32)

Characteristic features

1. It is a diffuse telangiectasia with *no swelling*, usually seen on the face, lips and buccal mucosa.
2. It takes the shape of a diffuse staining of purple or dark red discoloration flush with the skin, which disappears on pressure.
3. Present since birth, and it shows no tendency to progress or disappear throughout life.

Treatment

It is for removal of appearance causing disfigurement.

- In a girl: The blemish can be disguised by the skillful use of cosmetics.
- In a boy: Excision and skin grafting may be considered.



Fig. 2.32. Portwine stain over the right cheek. (Courtesy: Dr. U.S. Chatterjee, MS, MCh (Paed), MCh (Uro))

Naevus flammeus may be associated with specific syndrome like Sturge-Weber, Osler Rendu-Weber and Klippel Trenauny-Weber syndromes.

Sturge-Weber syndrome

Naevus flammeus is associated with a haemangioma of the ipsilateral cerebral hemisphere which may lead to Jacksonian epilepsy and mental retardation, glaucoma with loss of vision and buphthalmos.

Osler Rendu-Weber syndrome

Naevus flammeus is associated with haemangioma of the gastrointestinal tract, urinary tract, liver, spleen and brain.

Klippel Trenauny-Weber syndrome

Naevus flammeus is associated with osteohypertrophy of the extremities and AV fistula.

III. Strawberry angioma

Characteristic features

1. It is composed of immature vasoformative tissues.
2. Skin and subcutaneous tissues are often involved. Muscle and mucous membrane may also be involved. Submucous variety is prone to haemorrhage which sometimes becomes severe and alarming.
3. Presents with typical history.

The baby is normal at birth. The lesion appears as a red mark after birth between one and three weeks of age – this red mark increases in size rapidly by the age of a few weeks to 3 months – and ultimately develops into a strawberry-like swelling which protrudes from the skin surface.

It grows with the child up to 1 year and then it ceases to grow. Then afterwards the swelling may disappear by the age of 7 or 8 years, when involution is complete. (Strawberry is a fruit.)

4. Strawberry angioma can occur at any part of the body but are most seen on the head and neck.
5. Bright or dark red, raised, hemispherical swelling, usually 1 to 2 cm in size.
6. *The surface is irregular* and covered with smooth, pitted epithelium. There may be small areas of ulceration covered with scab.
7. The lesion is *soft and compressible*; *sign of emptying is present*.
8. The swelling is *not pulsatile* (cf. *aneurysm*).

Treatment

1. Better to wait up to the age of 7 or 8 years for natural involution to get a better cosmetic result.
2. The treatment is indicated when the swelling persists:
 - (a) *Conservative*:
 - (i) Application of carbon dioxide snow.
 - (ii) Injection of hot water, hypertonic saline, sclerosant fluid, or steroids.
 - (iii) Laser therapy using carbon dioxide or Nd:YAG laser.
 - (b) *Operative*: Excision with or without skin grafting.



Fig. 2.33. Typical strawberry haemangioma over the forehead. Red colour swelling with nodularity – looks like strawberry fruit. Always wait and watch till 7 years of age and assure the parents for spontaneous regression or resolution.

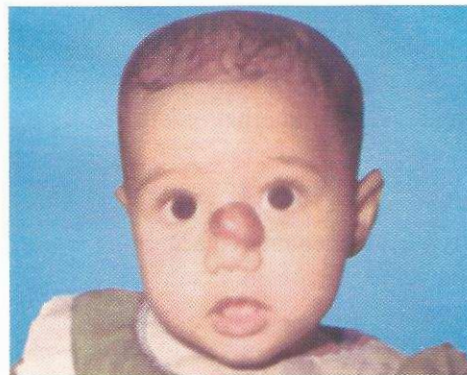


Fig. 2.34. Strawberry angioma over the nose. Usually the swelling disappears by the age of 7 or 8 years. So, better to wait up to that age for natural involution.



Fig. 2.35. Strawberry haemangioma on the right cheek. Note ulceration over the surface of the lesion.



Fig. 2.36A. Capillary haemangioma upper eyelid. Look other parts of the body, e.g., oral cavity.



Fig. 2.36B. Same patient. Also haemangioma noted in the palate and upper gum.

IV. Spider naevus

1. It is an acquired condition, mostly found in association with chronic liver disease (e.g., cirrhotic liver). It may occur as an isolated lesion.
2. It has a central red spot (a small dilated skin arteriole) with numerous linear, radiating, fine blood vessels like the legs of a spider.
3. Sign of emptying is present.

VENOUS OR CAVERNOUS HAEMANGIOMA

Characteristic features

1. This is due to the congenital multiplicity of venous channels having varying calibre.
2. Relatively uncommon, and present since birth.
3. Usually it shows no tendency to involution.

Rather it gradually increases in size and may become troublesome later.

4. It may be associated with a lipoma (naevo-lipoma). In some cases there may be presence of arteriovenous communications.

Common sites

1. Anywhere in the skin – commonly of the face, ear and the lip.
2. Subcutaneous tissue.
3. Mucous membrane of tongue, mouth, lips.
4. Internal organs – liver, kidney, brain, etc.

Diagnostic criteria

1. Presents as an appreciable swelling or as an area just raised from the surface.
2. Bluish discoloration.
3. Feel – soft, fluctuant swelling. May feel as spongy masses.
4. May be compressible. When compressible – *Emptying sign is present*. Compressibility may be absent (over the whole area or over some part) if there is associated thrombosis of underlying veins. This thrombosis may be spontaneous or due to the injection of sclerosing agent previously.
5. Not pulsatile – Pulsation in such a tumour indicates communication with arterial system.
6. Calcified nodules (phlebolith) can be palpated in rare cases.

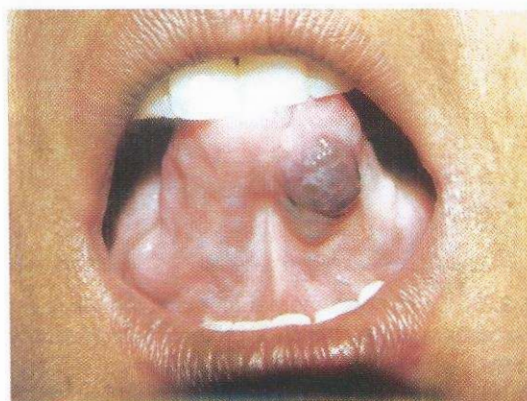


Fig. 2.37. Cavernous haemangioma undersurface of the tongue left side with bluish discoloration. Soft and compressible. Non-pulsatile. (Courtesy: Dr. S. Tibrewal, MS, MRCS, DNB, MNAMS)

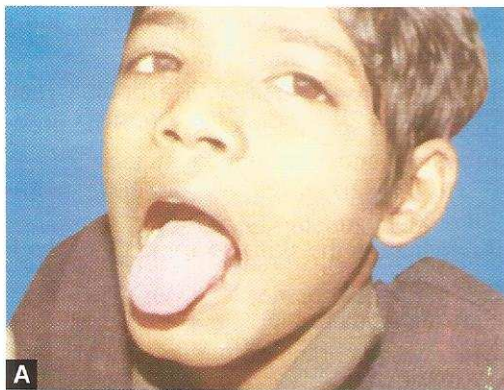


Fig. 2.38A. Cavernous haemangioma on the dorsum of the tongue at the middle – soft compressible swelling.

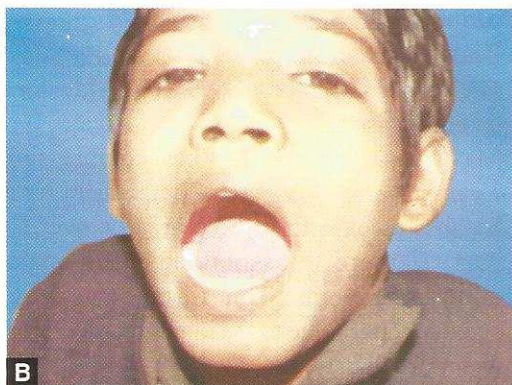


Fig. 2.38B. Same as above. Compression test positive.

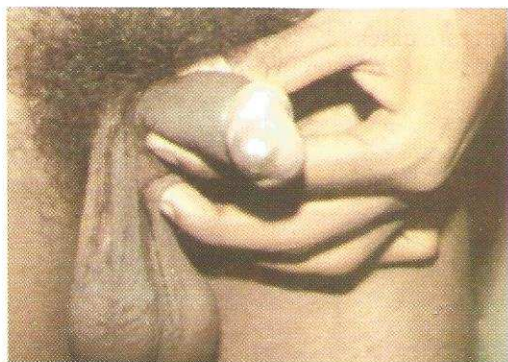


Fig. 2.39. Cavernous haemangioma at the glans penis. Soft, compressible, bluish swelling. Sign of emptying present.

ARTERIAL OR PLEXIFORM HAEMANGIOMA

This is really a type of congenital arteriovenous fistula where the arterial blood flows towards the vein and the veins become dilated, tortuous and thick-walled (i.e. arterialisation of the veins).

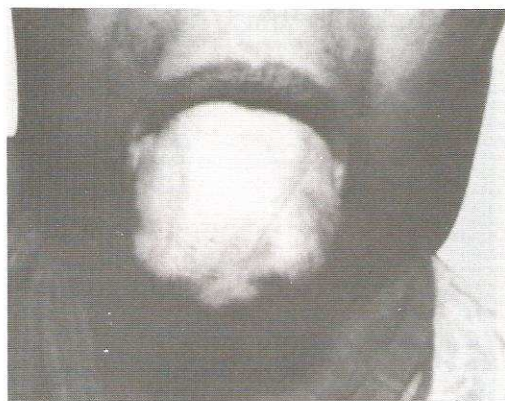


Fig. 2.40. Haemangioma of the tongue leading to macroglossia. (What are the other causes of macroglossia? – See page 145)

Example is *cirroid aneurysm* which is commonly found in the scalp (Fig. 2.42) – over the forehead and/or temporal region in relation to superficial temporal artery.

Diagnostic criteria

1. Diffuse swelling over the part: soft and cystic.
2. Feels like a bag of earthworms.
3. Swelling is pulsatile and compressible.
4. Systolic thrill and bruit are present.
5. When in relation to bone – X-ray may show evidence of osteoporosis or thinning out or destruction of the underlying bone.

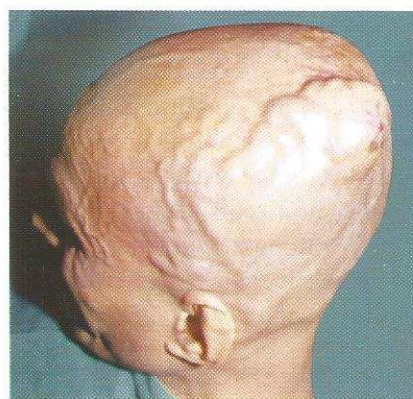


Fig. 2.41. Cirroid aneurysm over the scalp – left temporoparietooccipital region. Note the dilated tortuous veins over the swelling and also at periphery of the lesion. Excision of such lesion requires preliminary control and ligation of the feeding artery, e.g., superficial temporal artery and/or external carotid artery. (Courtesy: Dr. Sudeb Saha, MS, FAMS)

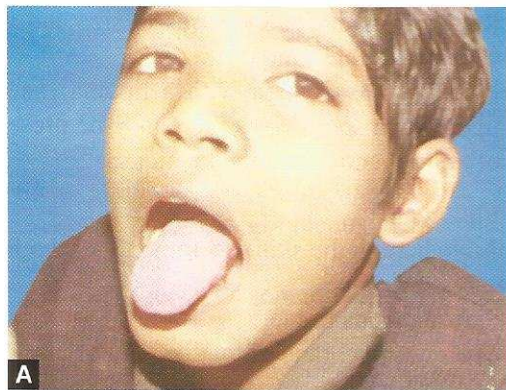


Fig. 2.38A. Cavernous haemangioma on the dorsum of the tongue at the middle – soft compressible swelling.

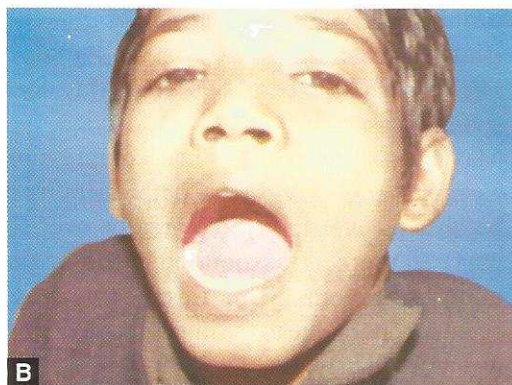


Fig. 2.38B. Same as above. Compression test positive.

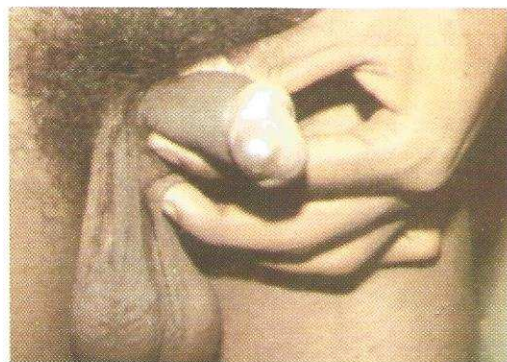


Fig. 2.39. Cavernous haemangioma at the glans penis. Soft, compressible, bluish swelling. Sign of emptying present.

ARTERIAL OR PLEXIFORM HAEMANGIOMA

This is really a type of congenital arteriovenous fistula where the arterial blood flows towards the vein and the veins become dilated, tortuous and thick-walled (i.e. arterialisisation of the veins).



Fig. 2.40. Haemangioma of the tongue leading to macroglossia. (What are the other causes of macroglossia? – See page 145)

Example is *cirroid aneurysm* which is commonly found in the scalp (Fig. 2.42) – over the forehead and/or temporal region in relation to superficial temporal artery.

Diagnostic criteria

1. Diffuse swelling over the part: soft and cystic.
2. Feels like a bag of earthworms.
3. Swelling is pulsatile and compressible.
4. Systolic thrill and bruit are present.
5. When in relation to bone – X-ray may show evidence of osteoporosis or thinning out or destruction of the underlying bone.

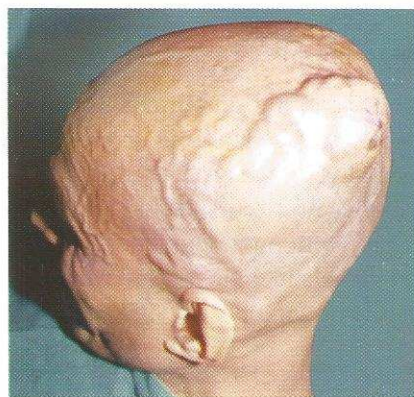


Fig. 2.41. Cirroid aneurysm over the scalp – left temporoparietooccipital region. Note the dilated tortuous veins over the swelling and also at periphery of the lesion. Excision of such lesion requires preliminary control and ligation of the feeding artery, e.g., superficial temporal artery and/or external carotid artery. (Courtesy: Dr. Sudeb Saha, MS, FAMS)

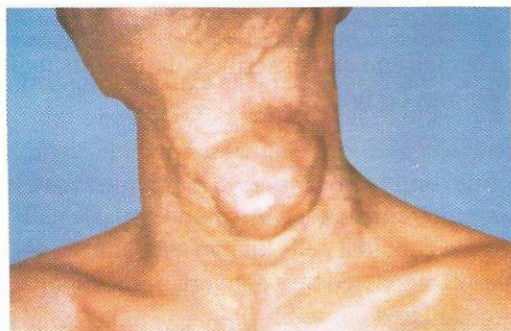


Fig. 2.42. Cirroid aneurysm over the neck. Soft swelling feels like a bag full of earthworms. Pulsatile and compressible. Thrill was present.



Fig. 2.44. This adult patient presented with *macrodactyly* of the left middle finger with extended broad linear swelling over the palm which was also extended to the wrist beneath the flexor retinaculum (note the ink marking of the swelling). The swelling clinically was felt like soft spongy mass and *compressible* throughout but *not pulsatile* (cf. cirroid aneurysm). There were no dilated veins visible over the dorsum of the hand. Cross fluctuation at the wrist was positive. Clinical diagnosis was haemangioma. Patient was advised (a) x-ray of the hand to look for bony changes, e.g., osteoporosis; and (b) colour duplex ultrasound of the hand to know exact cause of the swelling, but the patient did not turn up. Colour duplex ultrasound could be beneficial in the diagnosis of the lesion. This case will be difficult to treat; may require amputation of middle finger in addition to excision of the spongy mass. D/D: Compound palmar ganglion. (Courtesy: Dr. R.L. Modak, MS)

6. Duplex ultrasound is helpful in diagnosing and detecting the communication between the feeding artery and the veins.

Complications of haemangioma

1. Atrophy of the overlying skin.
2. Ulceration.
3. Haemorrhage.

4. Calcification.
5. Thrombosis.
6. Infection may lead to septicaemia.
7. Recurrence (Figs. 2.43A&B).
8. Pressure effects especially in skeletal haemangiomas: osteoporosis or bony erosion.
9. At times malignant change – haemangio-endothelioma, or haemangiosarcoma.
10. Diffuse haemangiomatous changes in a growing limb may cause its overgrowth.



Fig. 2.43A. Swelling on the right parotid region extending to the cheek with narrowing at the middle. Both were compressible. Scar mark from previous operation noted over the lower margin of the swelling – *Recurrent haemangioma*. Note: All the swellings over the parotid region are not parotid tumours.

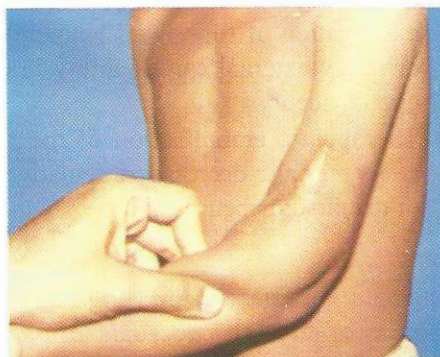


Fig. 2.43B. This patient presented with a diffuse, soft and partially compressible lump on the anterolateral aspect of the elbow extending to the lower 1/3rd of the arm to upper 1/3rd of the forearm. Note the hypertrophied scar from previous operation – *Recurrent haemangioma*. There were also some thickened nodules palpable which were due to thrombosis of the incompletely resected veins. Signs of blanching or surface discoloration absent – because of the nodularity. D/D: Neurofibroma.

General plan of treatment of haemangioma

I. Conservative

1. Wait and watch to expect spontaneous involution, e.g., in salmon patch, strawberry angioma.
2. Disguise the blemish (or colour) by skillful use of cosmetics, e.g., portwine stain, particularly in girls.
3. *Sclerotherapy*: Injection of hot water, or sclerosing agent into the haemangioma. Sclerosing agents are hypertonic saline, hypertonic glucose, etc.
4. *Steroid therapy*: Repeated injection of hydrocortisone into the lesion.
5. *Cryotherapy*: Application of carbon dioxide snow.

II. Surgery

1. If the lesion is localised:
 - Excision particularly by diathermy followed by skin grafting or extensive plastic procedures.
 - Sometimes excision is preceded by an injection of any sclerosing agent which will cause reduction in the size of the lesion.
2. If the lesion is extensive:
 - (a) Ligation of the feeding blood vessel preceding excision of the lesion may help in decreasing the size of the cirroid aneurysm, e.g. ligation of superficial temporal artery or rarely external carotid artery in case of cirroid aneurysm.
 - (b) Therapeutic embolisation of the feeding artery may also decrease particularly the size of the cirroid aneurysm.

III. Radiotherapy

Haemangioma is *radiosensitive*, but may result in disturbance of growth, necrosis of the skin, pigmentation, ulceration, haemorrhage, etc.

Radium mould sometimes helps, particularly when surgical access is difficult.

Treatment of cirroid aneurysm

1. Ligation of the feeding artery followed by excision of the mass.
2. Therapeutic embolisation of the feeding artery followed by excision of the mass.

GLOMANGIOMA (Syn. Glomus tumour)

It is a benign and circumscribed tumour arising from the 'glomus body'.

What is glomus body?

Glomus body is a specialized arteriovenous anastomosis surrounded by smooth muscle cells and large pale cuboidal cells known as glomus cells, the whole encompassed by a network of medullated and non-medullated nerve fibrils.

Common sites

Most abundantly present in the region of nail-bed, the tips of fingers and toes, and the palmar surface of the phalanges.

Function of glomus body

It regulates the temperature of the skin.

Characteristic features of glomangioma

1. Blue or reddish, circumscribed swelling, rarely exceeding 1 cm in diameter situated in the common sites as mentioned above. It is a benign tumour, grows very slowly and never becomes malignant.
2. *Severe pain*: The pain is burning or stabbing or lancinating in nature and radiates peripherally. The pain may be spontaneous or may be caused by the slightest pressure. The pain may be caused when the part is exposed to sudden change of temperature. *The pain is due to the pressure on the nerve endings by the dilated glomus vessels.*
3. On histological section: The tumour consists of a mixture of vascular spaces, nerve tissues and muscle fibres derived from the wall of the arteriole (angiomyoneuroma). There may be presence of large cuboidal cells (glomus cells), each with a central dark-staining nucleus.

Differential diagnosis

1. Subungual granuloma – chronic infective granuloma from lateral nail fold.
2. Subungual melanoma.
3. Sprouting granulation tissue – due to chronic osteomyelitis of the distal phalanx.
4. Squamous papilloma.
5. Granuloma pyogenicum.

Treatment

Excision of tumour is followed by immediate, complete and permanent relief.

LYMPHANGIOMA

Lymphangiomas, like haemangiomas, are congenital in origin. They are localised cluster of dilated lymph sacs in the skin and subcutaneous tissues *which fail to connect into the lymph system*.

The lymphangiomas are classified into three varieties:

- (a) *Lymphangioma simplex or circumscriptum*: This superficial variety arises from the cutaneous capillary lymphatics and presents as circumscribed lesion which appears as small blisters and slightly elevated skin patches.
- (b) *Cavernous lymphangioma*: Lymphangioma arising from the larger lymphatic channels.
- (c) *Cystic lymphangioma* (cystic hygroma): Multiloculated cystic lesions composed of many cysts of varying sizes ranging from a few millimetres to a few centimetres.

Characteristic features

1. Developmental anomaly; so, present and often visible at birth.
2. Sites: Usually found at the junction of limbs with the trunk and junction of neck with the trunk, i.e., around the shoulders, axillae, buttocks, groins and at the root of the neck.
3. Size: Lymphangioma circumscriptum appears as small multiple vesicles, whereas cystic hygroma appears as big cystic swelling.
4. Colour: Skin vesicles contain usually clear fluid and looks yellow or watery and translucent. When such vesicles contain blood they turn brown to black.
5. Consistence: Soft and spongy. Fluctuation can be elicited in big lesions, e.g., cystic hygroma.
6. *Transillumination*: Positive in big lesions (cystic hygroma).
7. Usually painless lesions. Occasionally the vesicles may be rubbed with the clothes and get infected and painful.
8. Regional lymph nodes usually do not enlarge until and unless the lesion gets infected.
9. *Treatment*: Excision is the treatment of choice.

NEUROMA

Tumour of the nerve is called neuroma. Neuroma is

of two varieties – true and false. True neuroma is rare.

According to the source of origin tumours of the nerves can be classified into two main groups:

1. Tumours arising in connection with the sympathetic system – these are called true neuromas and are very rare.
2. *Tumours arising in connection with the connective tissue of the nerve sheath – these are called false neuromas and neurofibroma is included in these groups.*

True neuromas again are of two varieties according to their development from the neural crest. The sympathetic system originates from the neural crest and develops along two lines:

- (a) *Chromaffin tissue*, which is mostly found in the adrenal medulla and may give rise to tumours known as *chromaffinomas* or *pheochromocytomas*.
- (b) *Primitive neuroblasts and adult sympathetic cells*.

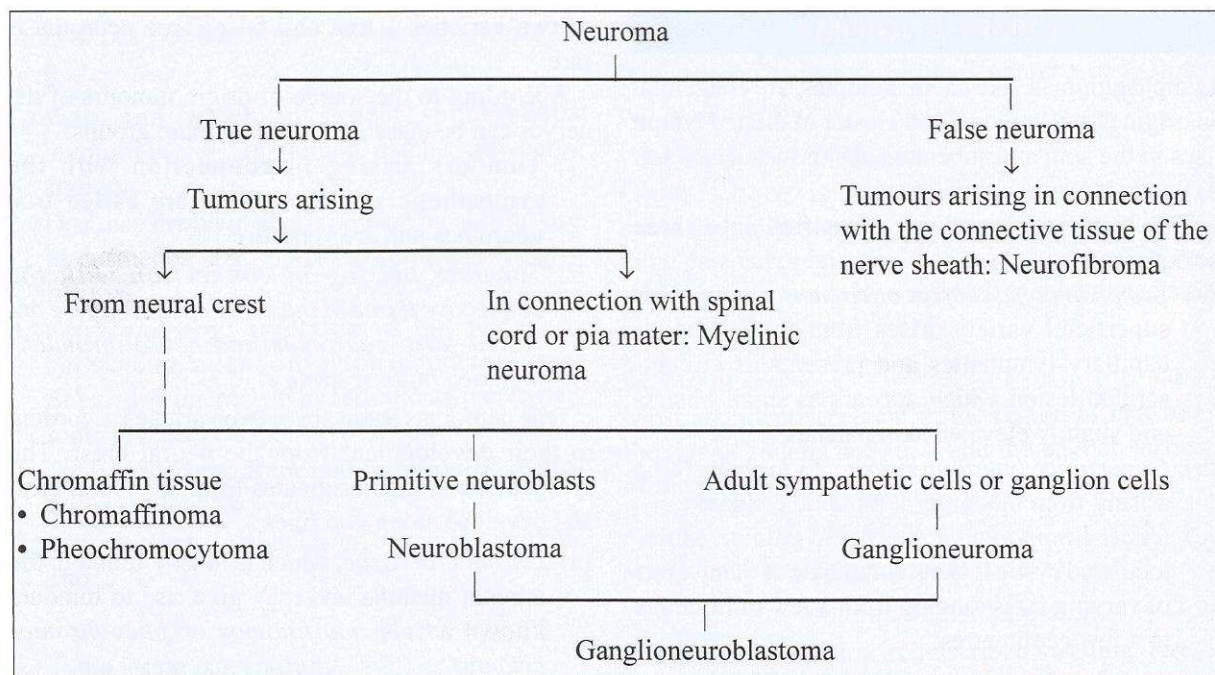
Primitive neuroblasts give rise to *neuroblastomas* which have the following features:

- Here the cells are of an embryonic type i.e., the tumours develop from unipotent cells.
- *Principally found in relation to adrenal medulla.*
- Usually occurring in infants and early childhood.
- *The tumour is usually malignant* and resembles a round-celled sarcoma.
- *Dissemination by blood is the rule.*
- Occasionally the tumour may undergo spontaneous remission.

Adult sympathetic cells or ganglion cells give rise to *ganglioneuromas* which have the following features:

- The tumour consists of ganglion cells, non-medullated nerve fibres and fibrous tissue.
- It arises in connection with the sympathetic cord and hence mostly found in the retroperitoneal tissue, or in the neck or thorax.
- It may occur at any age.
- *Relatively benign in nature.*

Both the neuroblastoma and ganglioneuroma are grouped together under the title 'ganglioneuroblastoma'.



There is another variety of true neuroma known as *myelinic neuroma* which consists of only nerve fibres; the ganglion cells are absent. The tumour arises in connection with the spinal cord or pia mater and is very rare.

Structures developing from the neural crest

- All the sympathochromaffin cells, i.e.
 - Chromaffin tissue
 - Primitive neuroblast
 - Ganglion cell or adult sympathetic cell
- Schwann's cell
- Suprarenal medulla
- Probably pia mater and arachnoid mater.

NEUROFIBROMA

Characteristic features

- Neurofibromas are not really tumours but regarded as *hamartomas*.
- These arise *not from the nerves proper* but from the *endoneurium* which is a supporting connective tissue for the nerve fibrils. The nerve fibres are not involved. The *epineurium* and *perineurium*, the outer layers of the nerve sheath, remain normal (cf. Schwannoma – derived from

the Schwann's cell intimately surrounding the axons).

- But neurofibroma can give rise to neurodeficits, sensory and/or motor, by causing compression of the nerve fibres
- Majority of neurofibromas arise from the peripheral nerves and are present in the skin and subcutaneous tissue.
- These are due to an autosomal gene defect and are transmitted as a 'Mendelian dominant'.
- These can appear at any age but usually present in adult life.

Types

- Localised neurofibroma or solitary neurofibroma.
- Generalised neurofibromatosis or von Recklinghausen's disease.
- Plexiform neurofibromatosis.
- Elephantiasis neurofibromatosa.
- Cutaneous neurofibromatosis of mollusum fibrosum.

SOLITARY NEUROFIBROMA

Pathology

- Occurring in connection with endoneurium.
- Usually found in the skin and subcutaneous tissue.

3. Usually it grows from a peripheral nerve, e.g., median or ulnar nerve commonly above the elbow, or cranial nerve, e.g. acoustic neuroma.
4. It forms an *encapsulated rounded swelling* lying alongside a nerve, often with the nerve fibres stretched over its surface. The swelling may resemble a fibroma.
5. Histologically long slender cells with elongated nuclei showing palisading arrangements are characteristic features.
6. It may undergo cystic degeneration or sarcomatous change.

Diagnostic criteria (Figs. 2.45A, B, C)

1. *Subcutaneous nodule*: Smooth, firm swelling in the skin and subcutaneous tissues in relation with the peripheral nerve.
2. Margins are well-defined.
3. Mobility: Mobile opposite to the axis of the nerve, i.e. in lateral directions.
4. Swelling sometimes becomes painful and tender.
5. There may be a history of paraesthesia or pain and/or weakness (rarely) of the muscles supplied by the nerve because of pressure of the tumour on the nerve fibres which are spread over the surface. (*So always examine the patient for the areas of sensation or for the muscle power supplied by the nerve.*)
6. It may be associated with other types of neurofibromatosis. (*So examine other parts of the body.*)

Sites other than skin where solitary neurofibroma can occur

- (a) Dorsal nerve roots and ganglion.
- (b) Intracranial nerve, commonly 8th cranial nerve – acoustic neuroma. May arise from 5th or 9th cranial nerves rarely.
- (c) Intramuscular.
- (d) Inside the bone.

Differential diagnosis

1. Fibroma.
2. Lipoma.
3. Neurofibroma sometimes undergoes cystic degeneration – so to be differentiated from a cyst.
4. It often simulates enlarged lymph nodes – particularly when present in the neck.

5. Haemangioma.

Complications

1. Neurological abnormalities such as paraesthesia and/or motor weakness in the area of nerve supply.
2. Cystic degeneration.
3. Sarcomatous change.
4. Infection.

Treatment

Complete excision of neurofibroma taking care so that the nerve is not injured. Under no circumstances partial excision is advisable as it encourages sarcomatous changes.

Often resection of the adjacent nerves along with the neurofibroma may be required which is followed by an end-to-end suture of the divided nerve.

Indication of operation

1. Cosmetically looks ugly.
2. Painful and tender neurofibromas.
3. Producing pressure symptoms by its position, e.g., causing unbearable pain or paraesthesia, motor weakness.
4. Showing evidence of sarcomatous change.

GENERALISED NEUROFIBROMATOSIS OR VON RECKLINGHAUSEN'S DISEASE

Pathology

1. Multiple and numerous neurofibroma, occurring in connection with the endoneurium.
2. This condition often runs in families. It is inherited as an autosomal dominant disease.
3. More common in males.
4. It is a genetic disorder.
5. It may diffusely involve the peripheral nerves, cranial nerves (e.g., acoustic neuroma) and/or spinal nerves (e.g., dumb-bell neuroma).
6. Sarcomatous changes may occur in 5 percent cases.

Diagnostic criteria (Figs. 2.45A, B & C)

1. Multiple nodules of varying sizes, from a pea to an orange, are scattered all over the skin surface – face, neck, trunk, and the limbs.
2. The nodules vary in consistency from soft to hard and may be sessile or pedunculated. In a typical case, the body surface has a 'cobble-stone' appearance.

NEUROFIBROMA

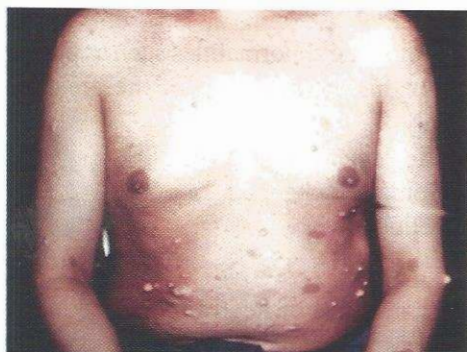


Fig. 2.45A. Multiple subcutaneous nodules over the abdomen and around both elbows – Neurofibroma.

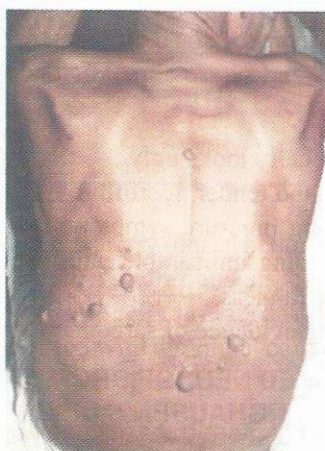


Fig. 2.45B. Multiple neurofibroma involving the back. The spine was straight. Examine the spine always for scoliosis or kyphosis as they are very often associated with neurofibromatosis. Look for cafe-au-lait spots also.

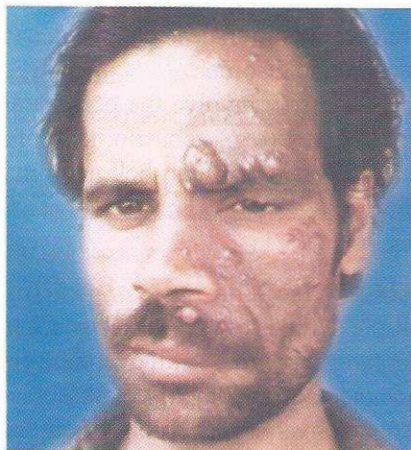


Fig. 2.45C. Multiple neurofibroma of the face. Note the pigmentation of the skin (cafe-au-lait spot). Look for lesions in any other part of the body, and any spinal deformity.



Fig. 2.46A. Plexiform neurofibroma over the right groin. Note the pigmented, thickened and corrugated skin folds. (Courtesy: Dr. Diptendra Sarkar, MS, FRCS, DNB)



Fig. 2.46B. Pachydermatocoele or plexiform neurofibroma involving the back of the lower 1/3rd of thigh and upper 2/3rds of the leg.

3. There may be associated pigmentations of the skin. These are *Cafe-au-lait spots* – the multiple patchy pigmentations in the skin or over the swelling, look like the colour of coffee diluted with the milk. The pigment is melanin. (The presence of skin pigmentation in some cases of von Recklinghausen's disease is a reminder of the common neuroectodermal origin of nerve sheath cells and melanocytes.)
4. It may be associated with the skeletal deformities namely kyphoscoliosis, deformities of the limb, osteoporosis of the bone. Examine the spine and the limbs.
5. Neurological abnormalities may be rarely present. So, look for it.
6. Sarcomatous changes may occur.

Treatment

The swellings are so numerous that excision of all the tumours is impossible and unwarranted.

Excision sometimes is indicated in the following conditions:

- (a) When one of the lumps becomes painful and/or tender.
- (b) When one causes pressure symptoms such as neurological abnormalities.
- (c) When one causes mechanical discomfort.
- (d) When one becomes large enough and unsightly.
- (e) When there is suspicion of malignant change.

Name any other disease under the name of von Recklinghausen's disease

von Recklinghausen's disease of bone or osteitis fibrosa cystica – found in hyperparathyroidism characterised by a parathyroid adenoma, bone resorption with or without pathological fracture, recurrent renal calculi and peptic ulcer.

PLEXIFORM NEUROFIBROMATOSIS OR PACHYDERMATOCELE (Figs. 2.46A, 2.46B)

Characteristic features

1. There is excessive overgrowth of endoneurium of the nerve in the subcutaneous tissue.
2. *Site*: Rare condition, usually occurring in connection with the branches of the trigeminal nerve (5th cranial nerve) in the distribution of face and scalp. It may occur in the extremities, arm or thigh along a group of nerves.
3. There is myxofibromatous degeneration of the endoneurium of the nerve and the affected nerve becomes enormously thickened.
4. Clinically, the overlying skin may be thickened, oedematous, pigmented and adherent, or it may be drawn out and folded. Sometimes the involved skin hangs down in pendulous folds. So, the patient usually presents with a mass hanging like a festoon which cosmetically looks ugly.
5. On palpation it resembles tortuous thrombosed veins due to the plexiform mass of thickened nerve fibres (cf. haemangioma).
6. It may be associated with von Recklinghausen's disease.
7. It may undergo sarcomatous change.

Differential diagnosis

From haemangioma particularly when thrombosed.

ELEPHANTIASIS NEUROFIBROMATOSA

Characteristic features

1. It is a severe form of plexiform neurofibromatosis.
2. A rare and congenital condition.
3. Limbs are common sites, particularly the lower limb and when the leg is affected, the patient finds walking increasingly difficult.
4. Clinically the skin becomes thickened, coarse and looks like an elephant's hide. The subcutaneous tissue become greatly thickened and the fat is replaced by fibrous tissue.
5. Usually it may be confused with the filarial elephantiasis but the lymphatics are normal.

Causes of elephantiasis of the limb

1. Filarial elephantiasis.
2. Arteriovenous fistula.
3. Nodular leprosy – Elephantiasis graecorum.
4. Occlusion of previously normal lymph nodes or lymph channels by:
 - (a) Surgical procedures (e.g., ilioinguinal node dissection).
 - (b) Malignancy.
 - (c) Radiotherapy.
 - (d) Inflammation.
5. Elephantiasis neurofibromatosa.

CUTANEOUS NEUROFIBROMATOSIS OR MOLLUSCUM FIBROSUM

Characteristic features

1. It occurs in connection with the terminal filament of cutaneous nerves.
2. The lesion appears as multiple cutaneous nodules particularly over the chest, abdomen or back.
3. The nodules are multiple, small, discrete, firm, sessile or pedunculated. But there is no hypertrophy of the skin (cf. plexiform variety).
4. Pigmentation of the skin may be present.

Complications of neurofibroma

1. By its position:
 - (a) Acoustic neurofibroma.
 - (b) Mediastinal syndrome when found in the mediastinum.
 - (c) Dumbbell-shaped neurofibroma.
 - (d) It may cause bizarre intestinal symptoms (abdominal neurofibroma).
2. Sarcomatous change.
3. Cystic degeneration.
4. Ugly deformity.

Clinical evidence of malignancy/sarcoma in a neurofibroma

1. Appearance of pain in a painless swelling.
2. Sudden increase in size.
3. Signs of paralysis or anaesthesia in the territory of the nerve distribution.
4. Evidence of increased vascularity.
5. Fixity to the surrounding structures.

RARE VARIETY OF NEUROMA

Amputation neuroma

Fusiform swelling occurs at the end of divided nerve after amputation of a limb. It is due to outgrowth of nerve fibrils which become attached to the skin, muscle or fibrous tissue.

This swelling consists of fibrous tissue and coiled nerve fibres. There is reparative proliferation of Schwann's cells.

These neuromas are painful which may be burning in character. There may be tingling and numbness on pressure over the neuroma.

Treatment

1. Preventive: During amputation, the nerves should be divided above the level of the proposed bone division.
2. Curative: Excision of the tumour.

Turban tumour

Form of neurofibroma arising from the cutaneous nerves of the scalp. Multiple neurofibromas may form multiple subcutaneous swellings which when coalescing may form a massive swelling which covers the head like a wig or turban, hence, the name 'turban' tumour.

Dumbbell-shaped tumour

It arises from the dorsal nerve root (lying partly outside and partly inside the intervertebral foramen giving an appearance of dumbbell). It causes root pain and paralysis.

Acoustic neuroma

It grows from the auditory nerve sheath at the internal auditory meatus.

As it arises from the 8th nerve, *the first symptom is unilateral deafness (perception deafness), which is often first detected by the patient on the telephone.* There may be tinnitus, vertigo and severe headache.

Gradually the tumour enlarges and projects into cerebellopontine angle and presses upon the adjacent nerves, e.g.

- (a) 7th nerve – causing facial weakness.
- (b) 6th nerve – causing squint.
- (c) 5th nerve – loss of corneal reflex; there may also be either trigeminal neuralgia or trigeminal anaesthesia.

If it becomes very large, the tumour may press upon the cerebellum and brain stem, producing cerebellar signs and raised intracranial pressure.

Treatment

Removal of the tumour through posterior fossa craniotomy.

Schwannoma (Neurilemmoma)

1. This is a benign tumour arising from the *Schwann's cells* which lies intimately surrounding the axon.

2. May be single or multiple and present as a fusiform swelling in relation with the nerves.
3. The swelling is soft, lobulated and well encapsulated.
4. Benign tumour and does not turn into malignancy.
5. It usually occurs from cutaneous nerve.
6. It can also be seen in mediastinum and retroperitoneum.

SCAR

It is a mass of devascularised fibrous tissues and it develops due to improper management of wound.

Complications

1. Contracture due to thickening and shortening of collagen bundles leading to the following defects:
 - (a) Deformity
 - (b) Limitation of movement.
2. Cosmetically ugly look.
3. Adherent scar which may cause either:
 - (a) painful movement or
 - (b) some limitation of movement.
4. Unstable scar or ulcerated scar due to:
 - (a) lack of nutrition due to avascularity, and
 - (b) lack of rest due to repeated movement.
5. Painful scar due to involvement of nerve in the scar tissue – commonly found in the scar of amputation stump.
6. Keloid.
7. Hypertrophic scar.
8. Marjolin's ulcer.

Management of scar

Preventive

1. *Infection of the wound should be avoided.*
2. Proper cleansing and dressing of the wound.
3. Excessive tension during the suturing of the wound edges should be avoided.
4. Early coverage of the raw area by proper skin grafting (as in case of burn). Ideal skin graft is split skin graft and such free graft should contain some amount of dermis.
5. By proper positioning of the wound.
6. When the scar is near the joint, the joint is maintained in a suitable position by proper splintage.

Curative

By operation.

Indication of operation:

1. Cosmetic (e.g., depressed scar as in smallpox).
2. If there is any complication as already mentioned.

Procedures:

The scar is excised and then the wound is covered by one of the following methods accordingly depending on the particular site of the scar and the nature of the defect.

1. Approximation of the edges after mobilisation of the skin margin. Excessive tension should be avoided.
2. Application of free skin graft.
3. By rotation flap.
4. By transposition of flap – the best and usual method is Z-plasty.
5. By pedicle flap.

Problems of operation:

1. The neurovascular bundle is frequently adherent to the scar tissue.
2. When the joint is deformed by the contraction of scar for a pretty long time the neurovascular bundle becomes short and contracted. So, during excision of the scar and correction of deformity the neurovascular bundle may be injured, or may be stretched or compressed.
3. Moreover, due to prolonged deformity there is secondary contracture of the joint capsule and the surrounding healthy tissue.

POST-BURN CONTRACTURE

It is contracture of scar developing after burn and occurring usually at the junctional areas, e.g. the flexor aspects of the body involving the nearby joint.

Causes

Post-burn contracture is a sequel to improper management of burn:

- (a) In case of deep burn involving the full thickness of skin where the subcutaneous tissue is exposed. So, healing does not take place by epithelialisation from the margins but occurs by fibrous tissue which when contracting leads to contracture.

- (b) In neglected cases of superficial burn which undergo infection and ulceration, the superficial burn is converted into a deep one and so heals by fibrosis. This fibrous tissue gradually matures and contracts leading to contracture.

Prevention of contracture

1. Proper treatment of superficial burns.
2. Adequate splintage of the part (e.g., a limb) in extension during the process of healing – to avoid development of contracture.
3. Prevention of infection: In the presence of infection the healing is delayed. Moreover, infection causes necrosis of the tissue which is then replaced by fibrosis, and thus adds to scar contracture.
4. Application of early skin grafting: The full thickness burn can never heal by marginal epithelialisation, so it must be skin grafted.

Early skin grafting prevents excessive scarring and so development of contractures. Moreover, early skin grafting prevents infection.

Common sites of post-burn contracture

At the junctional areas such as:

1. Neck
2. Axilla
3. Elbow
4. Groin
5. Knee

Complications

1. Keloid or hypertrophic scar
2. Deformity and limitation of movement
3. False ankylosis
4. Marjolin's ulcer
5. Cosmetically ugly look.

Treatment

Post-burn contracture should be released particularly to improve the deformity and to increase the movements of the adjacent parts at the junctional area. May require multistage operations. Also requires postoperative splintage to maintain correction.

Timing for release operations

The release operation is performed only after the scar tissue has matured. The scar maturation takes about 6 months to 1 year. Mature scar tissue becomes soft and supple in texture with a whitish hue.



Fig. 2.47. Post-burn contracture involving the right knee, ankle and lateral three toes. There is also hypertrophic scar over the dorsum of left foot.

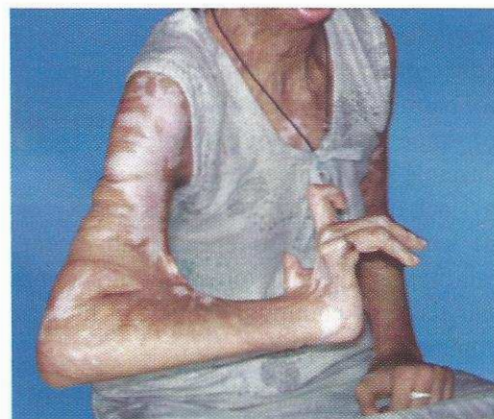


Fig. 2.48. Post-burn contracture involving right axilla, right elbow, right wrist, and right thumb and other fingers. This type of contractures require multistage operations.

Method of release

Usually by single or multiple Z-plasty at the point of maximum tension.

- *For neck scar contracture:* The fibrotic scar tissues are incised with multiple Z incisions at the point of maximum tension upto the healthy normal tissue from posterior border of one sternomastoid to the opposite sternomastoid. Incisions should be deepened upto the strap muscles of the neck. The neck is extended at its maximum and the raw surface should be covered with split skin graft

usually taken from the thigh. The neck is then splinted to maintain extension during the healing time.

Problem of anaesthesia in neck contracture

There may be great difficulty during endotracheal intubation which may be managed by one of the following methods:

- (a) Fibreoptic bronchoscope is used to allow endotracheal intubation.
 - (b) Blind nasotracheal intubation.
 - (c) Alternatively the neck contracture is released partially under local anaesthesia to allow nasotracheal or oral intubation.
- *For contractures of axilla or limbs:* The fibrotic scar tissues are incised by single or multiple Z incisions. The scar tissues are released, the joints are manipulated till the joint deformity is corrected. The raw surface after correction is covered with split thickness graft. The limbs are then splinted in corrected positions.

Postoperative splinting to maintain the correction

- Neck – Moulded cervical collar for 6 months.
- Axilla – Abduction splint for 3–6 months.
- Elbow and knee – Static extension splint (functional brace) for 3–6 months.
- Hand – Keep the hand in position of function in a glass-holding position for 2–3 months.
 - (i) Wrist – in 10–15 degree extension
 - (ii) Thumb – in palmar position
 - (iii) Fingers – Metacarpophalangeal joints at 80–90 degrees flexion and interphalangeal joints in extension.
- Postoperative physiotherapy under supervision particularly for limb contractures.

HYPERTROPHIC SCAR

It is a type of scar characterised by hypertrophy or proliferation of mature fibroblast or fibrous tissue without any proliferation of blood vessels (cf. keloid). It is caused due to some extra stimulus to fibrous tissue formation during healing such as excessive tension, infection, or foreign body response to a suture or other foreign material. Scars crossing the skin creases are particularly vulnerable to these complications.

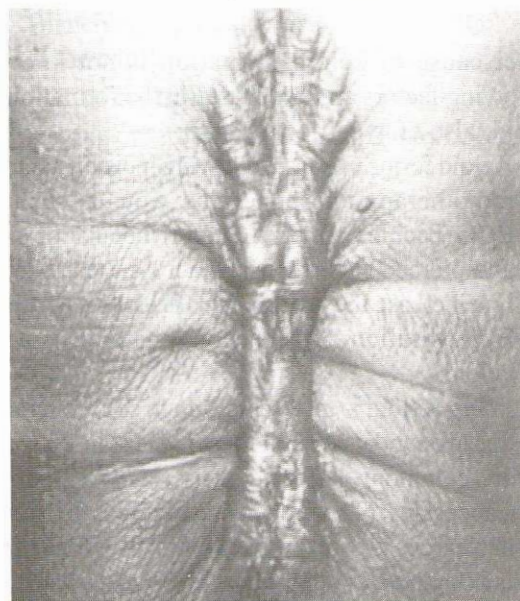


Fig. 2.49. Hypertrophied scar developing on the post-operative scar over the upper abdomen.

Characteristic features

1. The scar is raised above the surface.
2. Surface is glossy.
3. The hypertrophy is confined to the scar, i.e., *scar hypertrophy does not encroach areas beyond the dimensions of the original wound* (cf. keloid).
4. No claw-like processes.
5. No sign of increased vascularity.
6. No itching.
7. It does not spread.
8. The hypertrophic scar continues to enlarge for about 6 months but regresses after a year to pale, thin, stretched scar tissue. The hypertrophic scar never gets worse after 6 months (cf. keloid).
9. It does not recur after excision if the causative factors are eliminated.

KELOID

Keloid is a tumour-like lesion arising from the connective tissue elements of a scar and *grows beyond the dimensions of the original wound* (cf. hypertrophic scar).

It is characterised by disorderly proliferation of degraded collagen fibrils, immature fibroblast and immature blood vessels (cf. hypertrophic scar).

Aetiology

Exact cause of keloid formation is not known. Following factors are implicated in the formation of keloid (also of hypertrophic scar):

1. Keloid forms in a scar following incisions, burns, radiotherapy and punctured wounds such as BCG (bacille Calmette–Guérin) inoculation sites and pierced ears. Incisions across the Langer's line increase the probability of keloid formation.
2. Extravasation of sebaceous material – acne keloid.
3. *Race*: Keloid is more common in coloured races due to abundance of pigment in the skin. Negroes are commonly affected. It is least common in Caucasians. (The propensity of keloid formation in Negroes is exploited by Nilotic tribes of Africa for ornamental decoration.)
4. *Age*: Most common in young people aged 10–40 years. They do not form in the elderly.
5. *Sex*: Females are more commonly affected (more so in pregnancy).
6. Familial diathesis.
7. Hereditary influence.
8. Persons suffering from tuberculosis are more prone to suffer from keloid.

Sites of predilection

The most common site of keloid formation is on the sternum. Keloid also occurs, in decreasing frequency, on the shoulders, neck, face, extensor surfaces of the limbs and on the trunk. Keloid never occurs on the palms of the hands and on the soles of the feet.

Pathology

Due to irritation of the mesenchymal cells surrounding the base of the sebaceous and sweat glands immature fibroblasts proliferate in abundance together with the proliferation of collagen fibrils and immature blood vessels.

Due to proliferation of these cells microscopically it looks like soft fibroma, with the fibroblast disposed in parallel sheets embedded in the stroma of collagen.

Majority of the cells proliferate in claw-like processes to the surrounding tissue, hence the name keloid. Some keloids are, however, non-progressive.

Due to the presence of plenty immature blood vessels, blanching, oozing, itching and bluish discoloration of the keloid are commonly seen.



Fig. 2.50. Keloid involving the chest, neck, right shoulder and left arm. Also below and behind the right ear.

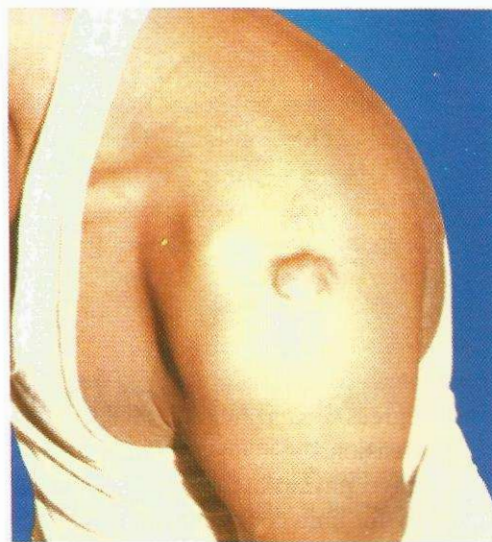


Fig. 2.51. Keloid developing on an injection site. D/D: Implanted dermoid.

Keloid can be considered tumour in the sense that new tissue is formed. But as there is no invasion or distant spread, keloid is not true tumour. However, it has a marked tendency to local recurrence after excision like a true tumour.

Types

1. According to the presence of capsule:
 - (a) Progressive: There are claw-like processes

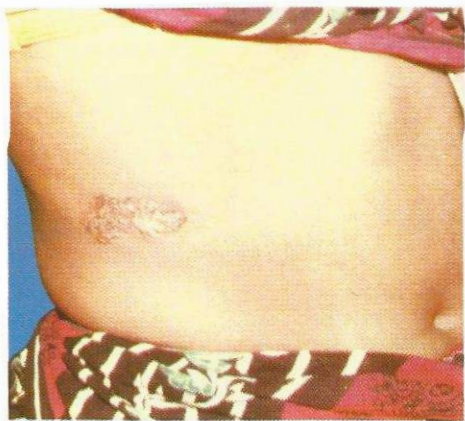


Fig. 2.52. Keloid developing over herpetic vesicles.

involving the greater area with more itching, oozing and blanching. Here there is no capsule. *The word keloid means claw-like process.* A true keloid continues to get worse even after a year, and some may even progress for 5 to 10 years (cf. hypertrophic scar – which never gets worse after 6 months).

- (b) Non-progressive: Usually surrounded by a capsule formed by the compression of surrounding connective tissue. This variety does not grow any further than a certain limit. So, claw-like extensions are not evident.
2. According to the mode of origin:
 - (a) Acquired variety: Developed after some sort of previous injury.
 - (b) Spontaneous variety: This variety grows spontaneously. Patient does not give any history of previous injury (such as pinprick, incision or burn). This variety usually occurs in the region of the chest and is like butterfly in shape. Spontaneous variety is commonly found in coloured races.

Diagnostic criteria

1. The swelling is raised from the surface.
2. Surface of the swelling is lobulated and firm in feel.
3. Usually there are claw-like processes and so the keloid extends beyond the original scar both in length and in breadth (cf. hypertrophic scar).
4. Bluish or pinkish in colour.

5. Blanching is present with presence of capillaries at the edge.
6. Patient complains of itching.
7. Oozing of serosanguineous fluid may be present.
8. It may be painful and tender.
9. Local temperature may be raised.

Differential diagnosis

Hypertrophic scar.

Complications

1. Cosmetically ugly look.
2. Recurrence of keloid.
3. Infection.
4. Suppuration.
5. Ulceration.
6. Malignant change on the top of keloid is called Marjolin's ulcer. (Some authorities believe that keloid can never undergo malignancy.)
7. If formed on the flexor aspect of a joint, keloid can lead to contracture and may interfere with mobility.
8. If located in a hairy area, there may be formation of pilonidal sinus which then enhances the further growth of the keloid.

Treatment

Treatment is very difficult.

Conservative

Usually it has no role, as the result is disappointing. Conservative treatment includes:

1. Intrakeloidal injection of hydrocortisone. The most effective agent is triamcinolone. The maximum dose is given – 0.5 ml at one site. The solution can be diluted and is injected with an insulin syringe. The injection is continued while withdrawing the needle. Administration should be weekly for 4–8 weeks.
2. Intrakeloidal injection of hyaluronidase – because it might help the dispersion of, or the solution of, the fibroblasts and collagen fibrils.
3. Intrakeloidal injection of vitamin A.
4. Parenteral administration of vitamin A.
5. Deep X-ray therapy. The ideas is:
 - (a) to suppress the proliferation of immature fibroblasts and collagen fibrils, and

- (b) to suppress the proliferation of immature blood vessels and thus to stop to a great extent itching and oozing.
- 6. Ultrasonic therapy – recently advocated for hypertrophic scar and keloid with a good result.

Operative

Established keloid (more than 6 month old).

Management:

Can be treated by excision.

1. Pre-operative radiotherapy (4 or 5 sittings are needed).
2. Excision of keloid: Incision is given in such a way as to leave a thin peripheral rim of keloid. The defect is closed by:
 - (a) approximation of the margins of the wound by suture, or
 - (b) application of split skin graft.
3. Post-operative radiotherapy – both to:
 - (a) the donor area and to
 - (b) the recipient area.

Idea of giving radiotherapy – to reduce the chances of recurrence.

PILONIDAL SINUS AND FISTULA

Pilonidal means a nest of hair (Latin *pilus*, hair; *nidus*, nest). The pilonidal sinus is a subcutaneous track containing hairs or their microscopic fragments.

Sites

Pilonidal sinuses are found most often in the natal cleft overlying the coccyx or lower part of the sacrum. They may, however, occur in other regions, e.g., interdigital clefts of barber's finger, the umbilicus, the axilla and the face.

Aetiology

Not definitely known and two theories have been suggested.

- (a) *Congenital theory:* It has been suggested that a developmental abnormality around the sacro-coccygeal region leads to the formation of an



Fig. 2.53. Pilonidal sinus at the upper end of natal cleft.

epithelial lined sinus which grows hairs at puberty. The lesion may be caused by any one of the following ways:

- The origin of the lesion is associated with the neuroenteric canal and is due to blockage of a congenital coccygeal sinus which is a remnant of this canal. But the squamous lining of the tract contradicts this view and suggests rather an ectodermal origin.
 - The lesion may occur as a result of sequestration or subcutaneous inclusion of epidermal structures due to the faulty fusion of the ectodermal lining at the cleft between the buttocks.
 - It may result from excessive traction on the skin caused by the retrogression of the tail bud.
- (b) *Acquired theory:* Recently this theory has won a major acceptance and claims that the sinus is nothing more than a foreign body granuloma, the foreign body being hairs which penetrate the skin through abrasions, sweat glands or hair follicles. The reasons which favour the theory of acquired origin of pilonidal sinus can be summarised as follows:
- The occurrence of pilonidal sinuses elsewhere – interdigital pilonidal sinus is an occupational disease of 'barbers' but the hair within the interdigital cleft or clefts is that of the customers.
 - Also the pilonidal sinus may occur in the umbilicus, the axilla or the face.
 - The lesion occurs most often in the late teens or early adult life and this age incidence is in dissonance with the age of onset of congenital lesions.

- The presence of a granulation tissue lining of the sinus track.
- The sinus is lined by skin.
- The absence of hair follicles and sebaceous glands in the walls of the sinus.
- The hair projecting from the sinus are dead hairs and their pointed ends are directed towards the blind end of the sinus.
- The lesion is mostly common in men; hairy men being most frequently affected.
- Recurrence is common in spite of adequate excision of the sinus track.

Mode of origin of acquired pilonidal sinus

It is due to entrapment in the natal cleft of shed hairs of local origin or from the nape of the neck and subsequent penetration of the same in this region either through the intact skin via the openings of sudoriferous glands or the raw skin due to fissure, abrasion, dermatitis or pustules. Shedding of hairs is much augmented by the shearing strain in the buttock when one sits on a hard seat particularly in a moving vehicle for a prolonged period and shearing of hairs, subsequent entrapment and penetration are much more frequent in those who use toilet paper after evacuation. Once a sinus has formed, there is possibility of other loose hairs being sucked into the pit by the intermittent negative pressure of the area.

The pilonidal sinus was common among the jeep drivers in army and has been referred to as "jeep-driver's disease" by army surgeons.

Pathology

The sinus extends into the subcutaneous planes as a bulbous diverticulum with or without branching side channels.

Lining of the tract

Usually stratified squamous epithelium.

Contents

1. Granulation tissue.
2. Epithelial scales and debris.
3. Hairs: These are dead hairs and are found either:
 - (a) lying loose in the sinus;
 - (b) embedded in the granulation tissue; or
 - (c) deep in the mature scar tissue.

Microscopically

Foreign body giant cells are commonly present.

Diagnostic criteria

1. Age: Common in late teens or early adult life.
2. Sex: Males are more commonly affected. Many of the patients are exceptionally hairy and are usually obese.
3. Ethnic group: More common in dark-haired *hirsute men*. The condition rarely occurs in *blondes*.
4. Occupation: Common in 'jeep drivers'. Barbers may present with pilonidal sinus in the webs between their fingers.
5. Presence of a sinus or sinuses in the midline in relation to the skin dimple at the tip of the coccyx, i.e., at the upper end of the cleft between the buttocks.
6. The sinus may have one or multiple openings and on probing the sinus passed upward and forward towards the sacrum.
7. Presence of a tuft of hairs projecting from its mouth.
8. The sinus may be discharging and the discharge from the sinus is either serosanguineous or purulent, and may be blood-stained. The discharge may be foul smelling due to sebum and may contain hairs.
9. Pain and/or tenderness at the bottom of the spine.
10. Occasionally the patient may present with an abscess when the mouths of the sinuses are blocked.

Complications

1. Recurrent inflammation.
2. Abscess formation.
3. Recurrence of sinus: Recurrence is probably due to the following reasons:
 - (a) Inadequate excision: A diverticulum of the main tract has been overlooked at the primary operation.
 - (b) Entry of new hair through the scar or skin.
 - (c) If the natal fold is deformed by scarring after operation, the least trauma may cause tearing of the scar, and the resulting cervices may be contaminated with coliform and cutaneous bacteria, which leads to the formation of a dermal abscess and later a sinus.

Differential diagnosis

1. Fistula-in-ano: In pilonidal sinus there is absence of an internal opening in the anal canal. The sinus

tract also passes upward and forward towards the sacrum and not towards the anal canal. In fistula-in-ano the opening is in between the tip of the coccyx and anus.

2. Sinuses secondary to suppuration of post-anal dermoid: Here the opening of the sinus is in front of the sacrum and coccyx and not behind. Rectal examination may reveal a cystic swelling in front of the sacrum.

Treatment

No single method of treatment is entirely satisfactory. The most effective treatment is complete excision of the sinus tracks together with all their ramifications.

Operative treatment should be carried out at a time when the infection is quiescent.

To facilitate the excision, methyl blue may be injected as a guide to determine the extent of arborization of the sinus tract.

Following excision of the sinus, closure of the wound may be accomplished either primarily or by secondary intention. So methods of treatment include:

- (a) *Excision and primary closure:* An elliptical incision is used and the sinus and its ramifications are excised along with all the debris and hair. The defect is then closed either by suture, split-skin graft or Z-plasty. The disadvantage of the primary closure is that there is a chance of haematoma formation and subsequent infection.
- (b) *Excision and closure by second intention:* Following excision, proper haemostasis is secured by diathermy and catgut ligature. The wound is then not sutured but kept open and packed by gauze and thus allowed to granulate and heal by second intention. This method is preferable in the presence of any infection.
- (c) *Laying open of the track and its ramifications* and removal of all debris and hair followed by suturing of the edges to the skin (thus marsupialization of the sinus). Free drainage is thus established. This procedure yields a good hairless scar.

Conservative approach of Maurice

The sinus tracks can be destroyed by careful installation of full strength pure phenol by means of

a syringe and needle until the tracks are full and overflowing. The area is then squeezed until all phenol and debris are extruded and a second injection is repeated, which is also squeezed out.

Treatment of pilonidal abscess

Incision and drainage is advised.

Post-operative measures

1. Avoid prolonged sitting, e.g., driving a car.
2. The site must be kept free of hairs in future to prevent further incursions of hair by shaving or by using a depilatory cream.

BASAL CELL CARCINOMA (Syn. Rodent ulcer)

It is a slowly growing skin tumour arising from the *basal cells* of the epidermis of the skin.

It may also arise from the hair follicles, sweat glands and sebaceous glands (pilosebaceous adnexa).

It may be single or multiple.

It is called '**rodent ulcer**' because the ulcer grows by burrowing the deeper tissues like a rat (rodent – a rat).

It is also called '**field-fire type of cancer**' because it is found in some cases that the ulcer tends to heal at one place while breaking out in the adjacent area, i.e., like field-fire, ulcers spread centrifugally.

Sites

1. It is one of the commonest tumours of the face. *Face is the common site* because it is exposed to ultraviolet rays of the sun. Majority of the lesions occur on the face above an imaginary line joining the angle of the mouth and the lobule of the ear.

So, the commonest sites are:

- on the nose;
- near the inner and outer canthi of the eye;
- on the cheek near the nasolabial fold;
- on the forehead, particularly in females.

It is also called '**tear cancer**' because it is commonly found along the region on the face where tears roll down.

2. Rarely it may occur on the other parts of the skin surface, i.e., on the other parts of the face, neck, scalp (specially the forehead and temple), pinna, very rarely on the trunk and limbs.

3. BCC at the centre of face, postauricular region, pinna and forehead are more prone to recur.
4. Very rarely it may occur on the squamous cell mucous membrane, e.g., tongue, oesophagus and anal canal.

Aetiopathology

1. It arises mainly in late middle life, i.e., at or above 40 years of age.
2. It affects males more than females.
3. It is prevalent in white-skinned people in tropical regions. So particularly prevalent in Australia and New Zealand.
4. It is a *very slowly-growing tumour* and often the lesion has been present for two or even three years before any treatment is sought. *It is of low grade malignancy.*
5. It occurs in those parts of the skin which are unprotected from bright sunlight of high actinic value, that is why face is common site. There is often an *occupational factor* – agricultural workers, fishermen and dedicated sunbathers being more prone to this lesion.
6. The tumour is frequently multiple. The multiple growths may be confined to one area, or they may occur in different areas.

The multiple basal cell carcinomas particularly develop in persons who have an arsenical dermatitis following the prolonged administration of arsenic. Arsenic was once commonly used in skin ointments.

Types

1. *Nodule*: Solid, non-fluctuant swelling but may look cystic.
2. *Plaque*: Firm, raised and red plaque.
3. *Ulcerative*.

Description of a typical lesion

At its inception it lies deep to the epidermis and at this stage it appears as a firm papule or a flat or slightly raised plaque, or a dark cystic swelling with or without tiny venules coursing across the surface.

Sooner or later the covering epidermis over the centre gives way and the growth takes the form of an ulcer with the following characteristic features:

1. *Edge of the ulcer* is raised and rounded but not everted. The rolled edge may be beaded with pearly opalescent nodules. Sometimes one edge

of the ulcer undergoes partial healing while it is extending at another edge for a time, but later breaks out again. Initially the shape is circular but as the growth spreads the shape of the ulcer becomes irregular.

2. *Floor of the ulcer* first superficial, then becomes deep and even proceeds up to bone. Central part of the ulcer may show temporary evidence of healing by formation of a scab, but this scab goes on forming and breaking down. The scab is formed of dried serum and epithelial cells. If it is picked off, the floor will bleed slightly. If the ulcer erodes deeper structures, the floor is covered with poor quality granulation tissue.
3. *Base of the ulcer* is indurated but less pronounced or barely perceptible, and may be fixed to the deeper structures.

Histopathology

It is composed of closely packed islands of uniform basophilic epithelial cells disposed in rounded masses or columns set in a stroma of cellular connective tissue.

Central cells are polyhedral containing large basophilic nucleus.

The peripheral cells are columnar, more deeply staining and have a palisade arrangements (i.e. perpendicular to the surrounding connective tissue).

Prickle cells and cell nests are absent.

Mitotic figures are usually absent. Stromal component is composed of chronic inflammatory cells and benign fibrovascular tissue.

Spread

- *Local spread*: By direct infiltration superficially, or deeply to involve the other structures (e.g., cartilage, bone) is *very common*.
- *Lymph spread*: Lymph node metastasis, either regional or distant, is *very rare* even in a growth of long standing.
- *Blood spread*: *Extremely rare*.

Investigations

- In a small tumour less than 1 cm in diameter – *excision biopsy* is preferred which provides both tissue diagnosis and cure at the same time.
- In a larger lesion – *wedge biopsy* from the edge of the tumour or ulcer to confirm the diagnosis.

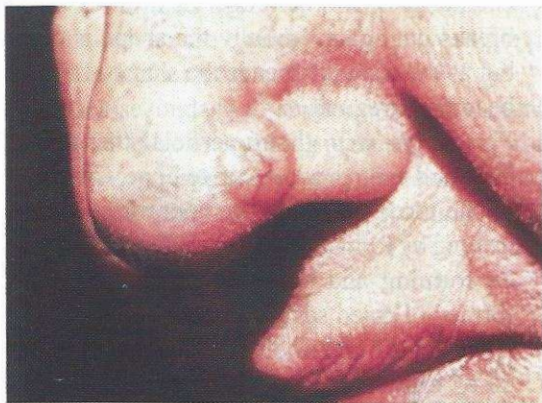
BASAL CELL CARCINOMA (B.C.C.)

Fig. 2.54A. Nodular type of B.C.C. on the left alae of the nose.



Fig. 2.54B. Plaque type of B.C.C. beside the right nasolabial fold.



Fig. 2.54C. Pigmented plaque type of B.C.C. just below the right lower eyelid. Note that all pigmented lesions are not melanoma.



Fig. 2.54D. Ulcerative type of B.C.C. on the left lower eyelid. Note the beaded appearances on the upper raised margin of the ulcer.



Fig. 2.54E. Ulcerative type of B.C.C. extensively involving the left eye. Both the burrowing (rodent) and spreading (field-fire) characters were evident in this lesion. (Courtesy: Dr. S. Tibrewal, MS, MRCS, DNB, MNAMS)

Treatment

1. Radiotherapy
2. Surgery
3. Other methods.

Radiotherapy:

- BCC is highly radiosensitive.
- Superficial radiotherapy using 40–60 Gy (4000–6000 rads) after preliminary biopsy and histological proof will cure over 90 percent of the lesions.
- The tumour may be so small that biopsy and cure are identical – the operation is called excision-biopsy. So, radiotherapy is avoided.

Contraindication of radiotherapy:

1. Adherence to bone or cartilage, e.g., over the nose or ear because of the high chance of subsequent necrosis.
2. Lesions very close to the eye to prevent radiation damage to eye.
3. Lesions on the back of the hand are also best avoided by the radiotherapist.
4. Exposure to extreme cold (as in Arctic regions) – as the exposed irradiation scars are very much prone to frost bite.

Surgery:*Indications:*

1. The causes where radiotherapy is contraindicated – as mentioned above.
2. Recurrence after radiotherapy.
3. Appearance of a new lesion closely adjacent to previously treated areas, either by radiotherapy or by surgery.

Principle:

Adequate excision of the lesion with a clear margin of *at least 3 to 5 mm and more* where possible. Adequate excision should also take account of the depth.

Reconstruction may be by simple direct suture or may need the use of split or full thickness skin grafts, to cover the defects.

In larger, neglected and deeply invasive lesions – plastic reconstruction by rotation or advancement flaps or pedicle grafts may be required, for better cosmetic results.

Other methods of treatment:

Recently, in some centres, the following methods are practised particularly in places where multiple tumours are common. The available methods are:

1. Curettage followed by diathermy.
2. Cryosurgery.
3. Laser beam destruction.

Disadvantages of these three methods are:

- (i) they destroy more tissues than are needed and thus result in retardation of healing;
 - (ii) they do not provide a tissue diagnosis;
 - (iii) not suitable for lesions situated over a bone or cartilage.
4. Local chemotherapy has been practised for some years in many centres – 5-fluorouracil cream is applied locally, and it is a simple and safe procedure particularly in flatter lesions.

BASISQUAMOUS CARCINOMA

It is a condition where histological features of squamous carcinomatous changes are seen often at the edge of a basal-cell carcinoma.

This type of lesion occurs more often in recurrent lesion or in the skin damaged by previous radiotherapy.

The lesion is more necrotic, friable and edge of the lesion is moist.

Lymphatic metastasis is a common possibility.

Diagnosis is confirmed only after biopsy.

Treatment should be done along the lines of squamous cell carcinoma.

SQUAMOUS CELL CARCINOMA (Syn. Epithelioma, Epidermoid carcinoma)

It is a malignant tumour arising from the squamous cells.

In the case of skin, it is the *prickle cell layer* from where squamous cell carcinoma arises.

Sites

1. *Any part of the skin or its appendages*, e.g., in face, dorsum of the hands, palm, sole, etc.
2. *Junctional region of the skin and mucous membrane*, e.g., lip, penis (corona glandis), anal region, vulva, etc.
3. Sites other than skin, e.g., mucous membrane:
 - (a) Mucous surface covered by stratified squamous epithelium, e.g.
 - (i) Upper air passage or food passage – tongue, buccal cavity, pharynx, larynx, oesophagus.
 - (ii) Vagina.
 - (b) Occasionally it may arise from the columnar cells undergoing metaplasia, e.g., in gall-bladder, bronchus, cardiac end of the stomach.
 - (c) Sometimes it may also arise from the transitional cell undergoing metaplasia, e.g., urinary bladder, pelvis of the kidney.

Aetiopathology

1. It usually occurs in elderly subjects.
2. It affects males more than females.
3. It is relatively common in white-skinned people in tropical regions.

4. It is more malignant and more rapidly growing than basal cell carcinoma.
5. Origin:
 - (a) *De novo*, i.e., it may arise in a previously normal area exposed to sunlight.
 - (b) *On some pre-existing skin lesion*: Pre-malignant lesions of the skin, e.g.
 - Senile (or solar) keratosis.
 - Bowen's disease – erythroplasia of Queyrat.
 - Leukoplakia.
 - Lupus vulgaris (a type of cutaneous tuberculosis).
 - Xeroderma pigmentosum.
 - Chronic ulcer of long-standing, e.g., venous ulcer, burns, old scar or keloid, chronic discharging osteomyelitic sinus causing chronic irritation (Marjolin's ulcer).
 - Radiodermatitis following irradiation.
 - Chronic irritation of the skin such as following:
 - Prolonged exposure to certain chemicals (carcinogens), e.g., pitch, tar, soot, dyes. Scrotal cancer was once common in workers of chimney sweeps (*chimney sweep cancer*). It still occurs in workers whose clothes become soaked in oil or pitch.
 - Continuous application of heat by a charcoal burner (Kangri) to the abdomen or back of the thighs may cause typical *Kangri cancer of Kashmir*.
 - Sleeping on oven bed, which is often a habit of Tibetans, may cause *Kang cancer of buttocks, heels and elbows*.
 - Intensive use of arsenical preparations such as Fowler's solution for psoriasis.

Progress

A squamous cell carcinoma may be invasive *ab initio*. Occasionally, however, dysplastic changes occur in skin epidermal cells, which later progress and present with all the features of malignancy with the exception of invasion. This later condition may involve any squamous epithelium, but is particularly common in the larynx or cervix and is called carcinoma-in-situ

or intraepidermal carcinoma. The latent period to the development of invasive cancer in such patients appears to be of the order of 10–20 years.



Fig. 2.55. Squamous cell carcinoma over the occipital region of the scalp – proliferative type with surface ulceration.

Types

1. Proliferative type or cauliflower type.
2. Ulcerative type – common

Description of a typical ulcerative lesion such as occurs on the lip (see also chapter on carcinoma tongue and lip)

To start with, there is a slight thickening or a small nodule which breaks down to form an ulcer. *This ulcer refuses to heal and is covered with a crust.* Then the ulcer becomes irregular and has the following characteristics:

- *Edge of the ulcer* is rolled out and everted.
- *Floor of the ulcer* is covered by greyish white slough.
- *Base of the ulcer* is indurated and may be fixed to the deeper structures.

Histopathology

It is composed of solid, irregular strands and columns of invading epithelium which infiltrate the subjacent connective tissue. In a well-differentiated case there is the presence of “cell nests” or *epithelial pearls of central keratin* surrounded by large peripheral eosinophilic or prickly cells in a concentric manner. These cells may show signs of malignancy (hyperchromatic nuclei, loss of polarity, mitotic figures).

What are "cell-nests"?

Epithelial cells of epidermis proliferate into the dermis in columns separated from one another by the connective tissue. In course of time the central cells, being the oldest, undergo some degenerative changes as found in normal surface epithelium and are finally converted into a hyaline structureless mass of keratin – *cell nests* (looks red after eosin stain). This process is called keratinization or cornification. This is surrounded by peripheral cells in a concentric manner giving an *onion-peel appearance*.

Peculiarities of cell-nests

- Well formed cell-nests in a squamous cell carcinoma indicate:
 - a relatively slowly growing tumour;
 - high degree of cell differentiation;
 - poor sensitivity to radiotherapy.
- Squamous cell carcinoma without cell-nests, but consisting of ill-differentiated or undifferentiated malignant cells, indicates:
 - a rapidly growing tumour;
 - sensitive to radiotherapy.
- Cell nests may be absent in a squamous cell carcinoma, e.g.
 - in a rapidly growing or well advanced tumour;
 - in the oesophagus and bladder, where cornification does not normally occur.
- Cell nests may be found rarely in some lesions other than squamous cell carcinoma, e.g.
 - pleomorphic adenoma of the parotid gland;
 - teratoma of the testis.

Spread

Local spread – by continuity and contiguity into the surrounding tissues is generally slow.

Lymph spread – by permeation and embolism, usually in the late stage. Lymph node involvement varies with the site of the primary lesion:

- It is common in cancer of the foot.
- It is frequent in cancer of the face or neck.
- It is late or absent in case of the hand of an old person, in case of a scar, or chronic ulcer.
- It is absent in Marjolin's ulcer.

Blood spread – occurs only in very advanced stages.

Investigation

To confirm diagnosis, an incisional or wedge biopsy is taken from the edge of the ulcer or tumour i.e., from the junction of the tumour and normal skin.

The reasons for taking biopsy from the edge are:

- The edge is the growing part, so malignant cells are plenty.
- Biopsy from the centre of the lesion may show only necrotic tissue.
- Comparison with the normal skin is possible.

For lesions ≤ 1 cm in diameter excisional biopsy is preferred which provides both tissue diagnosis and cure at the same time.

Treatment

- Treatment of the primary lesion.
- Treatment of the secondary lymph nodes.

Treatment of the primary lesion

Radiotherapy

- Superficial radiotherapy after adequate biopsy and histological proof will cure over 80 percent of early lesions.
- Radiotherapy is applied in the form of deep X-ray therapy, electron beams or local methods such as radium implants and moulds.
- *Radiotherapy is indicated in early lesion* i.e., well-differentiated tumour with margins free of tumour.
- *Radiotherapy is contraindicated* in certain areas such as:
 - Pinna – where radiation necrosis is a frequent and painful sequel.
 - Scalp – where radiation causes wide depilation, so this area is a cosmetic indication for surgery.
 - Lesion close to the eye.

Surgery

Indication

- Lesions are large in size.
- Involvement of the muscle, cartilage or bone.
- Lesion close to the eye.
- Lesion in the scalp, pinna.
- Recurrence after radiotherapy.
- Meagre facilities of radiotherapy, particularly in our country.

Principle

1. *Wide excision of the growth* with a margin of 1 to 2 cm of normal healthy tissue from the probable indurated edges of the tumour upto the deep fascia.
Reconstruction – The resulting defect may be closed by simple direct suture of the apposed skin margins. If the defect after excision is large, split or full thickness graft may be required to cover the defect. Rotational flaps or pedicle grafts may be tried for better cosmetic result.
2. Amputations may be necessary in some sites e.g., carcinoma of penis, or carcinoma of foot involving bones and tendons.
3. Lesions of the ear are treated with wedge excision when possible.

Treatment of the secondary lymph nodes

1. *Clinically no palpable nodes*: Regular observation is needed. If nodes appear, radical block dissection is to be performed after biopsy.
2. *Lymph nodes are palpable but discrete and mobile*: It may be reactive due to infection rather than involved by metastasis.
So, a course of antibiotics is given preoperatively for three weeks, and observe:
 - *If the nodes disappear or diminish in size* – suggestive of infective origin. So, no further treatment, but regular observation is required.
 - *If the nodes remain unaltered* – suggestive of malignant involvement. So radical block dissection is to be performed after biopsy.
3. *Lymph nodes are palpable, hard, mobile* and are suspected of being involved due to metastasis – radical block dissection is to be performed after biopsy.
4. *Lymph nodes are palpable, hard and fixed* – palliative radiotherapy is given.

MARJOLIN'S ULCER

It is actually a low-grade epidermoid or squamous cell carcinoma arising from the epithelium covering the scar tissue or keloid.

It may also develop on long-standing venous ulcer, long-standing radiation ulcer, long-standing ulcer following thermal injury, or on chronically discharging osteomyelitic sinus.

As the epithelium covering the scar tissues contains no hair follicle, no nerves, no lymphatics, the lesion has the following characteristic features.

Characteristic features (Figs. 2.56A, B, C)

1. It may occur many years after the formation of scar.
2. Very slowly growing – due to relative avascularity of the scar tissue. It is also less malignant than a typical squamous cell carcinoma.
3. Painless – as the scar tissue contains no nerve.
4. No lymphatic metastasis – as the lymphatics have been destroyed and occluded, unless the normal healthy skin adjacent to the lesion is involved when it presents the features of ordinary squamous cell carcinoma.
5. Radio-resistant – due to avascularity. Usual squamous cell carcinoma is radio-sensitive.

Treatment

After confirmation by biopsy:

1. If the site permits – wide tri-dimensional excision with a margin of at least 1 cm is performed followed by skin grafting, either by split skin or full-thickness skin.
2. If the ulcer is very big one involving the distal part of the limb and excision is not possible – the treatment is amputation.
3. Radiotherapy has no role.

MELANOCYTIC TUMOURS AND MALIGNANT MELANOMA

Melanocytic tumours are the melanin-containing tumours arising from the melanocytes or melanoblasts.

Surgical physiology

MELANIN: It is a sulphur-containing, iron-free black pigment. It is an endogenous pigment manufactured by specialised cells (melanocytes or melanoblasts). This naturally occurring pigment is normally found in the skin and choroid of the eye. It is also present in the following sites:

- (i) leptomeninges
- (ii) adrenal medulla
- (iii) certain nerve cells e.g., substantia nigra and locus ceruleus
- (iv) telachoroidea.

MARJOLIN'S ULCER



Fig. 2.56A. Marjolin's ulcer – squamous cell carcinoma of right forearm. Ulcer developed over the post-burn wound. Note the surrounding scar following burn.



Fig. 2.56B. Marjolin's ulcer (S.C.C.) on the medial side of the right leg. Note the hypopigmented surrounding scar. Required wide excision and skin grafting. (Courtesy: Dr. Sudeb Saha, MS, FAMS)



Fig. 2.56C. Clinically Marjolin's ulcer of the left leg. Note the hypertrophied scar above the ulcer. But after excision, histopathology of the lesion revealed schwannoma. Patient was treated with wide excision of the lesion followed by delayed skin grafting. Patient also had a course of post-operative radiotherapy.

Limited pigmentation may also occur in mucocutaneous junction, e.g., mouth, anorectal junction, vagina, nail bed, iris, ciliary body, etc.

The function of melanin is protective, and in human the melanin of the skin appears to protect it against sunlight.

Site of melanin formation

In the skin, melanin is formed in melanocytes or their precursors, the melanoblasts. Regarding the origin of these cells there are two views:

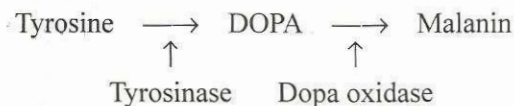
1. Epithelial theory: The melanocytes are specialised epithelial cells derived from the surface epithelium.
2. Neurogenic theory: The melanocytes are believed to originate from the neuroectoderm of

the embryonic neural crest and to migrate with the peripheral nerves to the basal layers of the epidermis (Masson's hypothesis).

Formation of melanin

The melanin is formed from the amino acid 'tyrosine'. Tyrosine is converted (oxidised) into dihydroxyphenylalanine (DOPA) by the enzyme tyrosinase which is present in the melanocytes. This DOPA is ultimately transformed into melanin after several stages by the action of an enzyme called 'dopa-oxidase'.

Melanin is intracellular and found near one of the poles of the cells as brown or black granules of uniform size. There are two varieties of cells – melanocytes and melanophores.

DOPA reaction:

The melanocytes are the mature cells. They contain the pigment melanin and also the enzyme 'dopa-oxidase'. So the functions of melanocytes are both synthesis and carriage of melanin. Melanocytes are dopa-positive. *Normally the melanocytes appear as clear cells in the epidermis, but in melanotic condition they are increased in number with pigmentation.*

The melanophores are phagocytes. They only carry the melanin pigment but cannot produce it as they do not possess the enzyme dopa-oxidase. So, the *melanophores are dopa-negative.*

Role of hormone

Synthesis of melanin and its progress are hormone dependent. Hormones responsible are:

1. Melanocyte-stimulating hormone (MSH) which is formed in the anterior lobe of the pituitary.
2. ACTH – to a slight extent.
3. Sex hormones:
 - Oestrogen
 - Androgen.

Surgical pathology

Melanocytic tumour may be benign or malignant.

BENIGN MELANOMA

(Syn. Melanocytic naevus, Pigmented naevus, Mole)

Naevus is a general term referring to localised cutaneous malformations including moles and birth marks. *Naevus means a birth mark.*

Melanocytic or pigmented naevi are those which result from the proliferation of melanocytes – pigment containing cells. *In the normal skin melanocytes are present in the basal layers of the epidermis.*

Pigmented naevi are common in all races and may be several in numbers. They may arise on any part of the skin surface and mucocutaneous junction of the body. They occur on the soles in about 10–15% of individuals.

Pigmented naevi may be present at birth, but more commonly they develop in childhood and at puberty or even later.

The following clinicopathological varieties of pigmented naevi are encountered:

- Congenital melanocytic naevi
- Acquired melanocytic naevi
 - (i) Junctional naevus
 - (ii) Compound naevus
 - (iii) Intradermal naevus

Congenital melanocytic naevus

- Appears at birth
- Carries a risk of malignant change.

Acquired melanocytic naevus

- Appear after first 6–12 months of age.
- Enlarges with body growths and may regress in later life.
- *In the normal skin, the melanocytes are present at the basal layers of the epidermis.*
- Initially, the naevus is junctional where proliferating melanocytes (naevus cells) are present in clumps at the epidermo dermal (ED) junction – *junctional naevus.*
Over a period of time, some of the melanocytes enter the dermis resulting in a *compound naevus* – the naevus cells being then present both at the epidermo-dermal junction and in the dermis.
- Later an *intradermal naevus* is developed where the naevus cells are present only in the dermis.

It is the junctional component of a naevus which is potentially dangerous, because it is these cells which are liable to undergo proliferation from time to time. *This is called junctional activity.* If this activity occurs in a naevus of an adult over the age of 30 years, the matter is serious, for it may be the first indication of malignancy.

Characteristic features of each variety**Junctional naevus**

- Most acquired melanocytic naevi in children appearing before puberty are of junctional type.
- Usually appear as circular or oval macular lesion that are smooth, flat or slightly raised, 1 mm to 1 cm in diameter.
- Shows variation of colour even in a single lesion – light brown to brownish black, often darker at the centre than at the periphery.
- Usually hairless.

- Can occur anywhere on the body and at any time after birth to puberty.
- *Most pigmented naevi on the soles, palms, digits and genitals are of junctional type.*
- Microscopically there is proliferation of melanocytes at the epidermodermal (E.D.) junction with varying amounts of melanin. These cells at E.D. junction are potentially unstable and may become malignant.
- *It is presumed that majority of malignant melanomas (90 percent) begin in junctional naevi, but all junctional naevi do not change into malignancy.*
- Some pathologists believe that malignant melanoma can also develop into compound naevi.

Compound naevus

- Combination of both junctional and intradermal components.
- The intradermal components are inactive and incapable of multiplication or pigment production. *But the junctional components are very much prone to malignant change.*
- More common in older children and adults.
- Appear as dome shaped pigmented nodules.
- Colour varies from brown to black. The centre is usually darker than periphery.
- Most lesions have a smooth surface but larger ones may have papillomatosis, hyperkeratosis. Many lesions may bear hair.
- *Junctional naevus and compound naevus are definitely pre malignant.*

Intradermal naevus (Common Mole)

- Found in adult and elderly subjects.
- Appear as an elevated, dome shaped, soft, smooth or warty lesion.
- Skin colour is light brown with or without telangiectasia.
- Mostly hairy, hence popularly known as *hairy mole*.
- Lesion can occur at any part of body except the soles, palms or genitalia. Mostly common on the face.

Clinical presentation of the benign pigmented mole

- Congenital pigmented naevus

- Multiple pigmented naevi
- Spitz naevus
- Blue naevus
- Dysplastic naevi
- Freckle

Congenital melanocytic naevi

- *Derived from epidermal melanocytes.* These naevus cells have a predilection for deeper infiltration.
- *Appears at birth.*
- Naevi may be *single* or *multiple*.
- Sizes vary from small to large: small lesion – less than 1.5 cm, intermediate lesion 1.5 cm to 20 cm. *Giant lesion* – more than 20 cm. Giant lesion usually seen on the trunk and because they may cover large areas of the trunk, they are called *bathing trunk naevi*.
- Colour varies from brown to black.
- The lesions darken and enlarge as the child grows.
- With age, the lesions also become elevated and develop rugosities.
- Coarse hair develops over most of the lesions – *Giant hairy mole*.
- Larger lesions may have satellite papules at the periphery.
- *Carries a definite risk of malignant transformation more so with giant hairy mole.*
- Spina bifida, meningeal involvement seen in lesions over the vertebral column.

Multiple pigmented naevi

- *Usually appear after puberty.*
- Commonly appear on the face.
- Women are more affected.
- They may become quite large and ugly.
- Microscopically, the cells are all in the dermis and no cellular activity is seen.
- *Malignant change usually does not occur.*

Spitz Naevus (Syn. Juvenile melanoma)

- *It is a type of compound naevus.*
- Predominantly seen in young children – so, it is called juvenile melanoma.
- Commonly appears on the face and lower extremities.
- Appears as a solitary erythematous nodule which grows rapidly over a period of one to two months and then become stationary.

MELANOMA

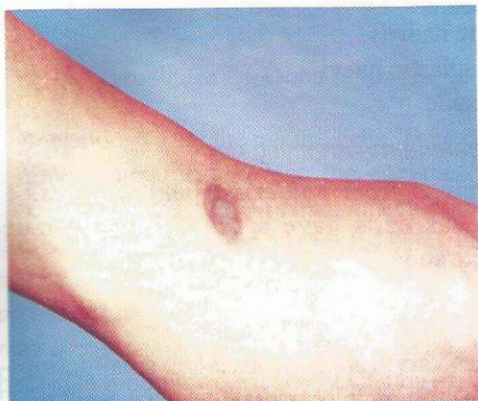


Fig. 2.57A. Hairy mole over the dorsum of the right hand.



Fig. 2.57B. Giant hairy mole right forearm.

- Benign lesion. But the lesion is often have to be excised *because of suspicion when there is rapid growth.*

Blue Naevus

- It is a special type of intradermal naevus.
- Microscopically, the cells are elongated, star-shaped having dendrites. The cells are melanin containing and situated deep in the dermis in strap like manner or in whorls. *The epidermis itself is normal.*
- Seen commonly in Mongoloid and Negroid infants.
- Appears as a macule or slightly raised, hairless lesion.
- Surface is smooth and shiny.
- Colour is uniform and varies from blue to bluish black.
- Commonly seen on the face, dorsum of the foot and hand, and buttock of the baby (Mongolian spot).

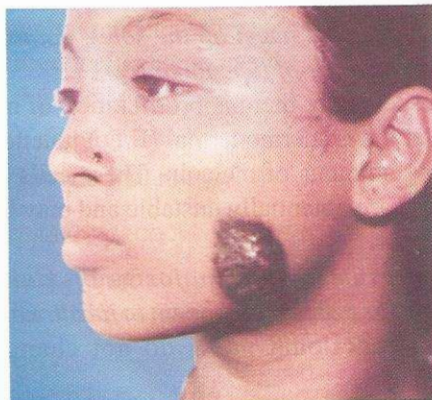


Fig. 2.57C. Darkly pigmented hairy mole over the left side of face; recently being enlarged, thickened and darker. Suspect malignant change.



Fig. 2.57D. Pigmented plaque type of lesion on right side of the face over the angle of the mandible since birth. Recently increasing in size during last six months with the development of satellitosis at the lower pole. Note that the lesion is also situated in an area subjected to repeated trauma from shaving. Total excision biopsy of the lesion including satellitosis and a margin of 2 mm of normal surrounding skin beyond the lesion was performed. Fortunately, histopathology did not show any malignant change.

- It is benign lesion, but very rarely may undergo malignant change.

Dysplastic Naevus

- Appear as multiple, large, irregularly hyper-pigmented papules (i.e., slightly raised from the surface); some having an erythematous halo.
- Common over the trunk; may appear over the scalp.
- Can occur sporadically, but *may be inherited as*

an autosomal dominant trait with incomplete penetrance – Dysplastic Naevus Syndrome; other family members may have such lesion.

- *Risk of malignancy is high*, so patient and other family members with dysplastic naevus syndrome should be followed up carefully.

Freckle (Syn.: Hutchinson's melanotic freckle (HMF), or Lentigo maligna)

- This condition starts as dark pigmented, smooth but flat lesion, i.e., macule.
- It commonly appears on the face and neck of elderly persons. The oral mucosa and conjunctiva can be involved.
- It is clinically a pre-melanomatous dermatosis but histologically is a *melanoma-in-situ with junctional changes* but no dermal invasion.
- It deserves special interest for three reasons:
 - (a) its late development;
 - (b) its tendency for steady centrifugal enlargement;
 - (c) its high incidence of malignant change – there may be raised rough nodules somewhere in the lesion.

Clinical evidence of malignant change in a naevus

1. *Increase in size.*
2. *Change in thickness:* Thicker, more palpable and nodular.
3. *Change in outline:* The edges become irregular.
4. *Change in colour:* Increasing pigmentation, depigmentation, irregular pigmentation.
5. *Change in surface characteristics:* Scaling, crusting, erosion, ulceration, oozing, bleeding, loss of hair from a previously hairy lesion.
6. *Change in surrounding tissues:* Appearance of pigmented halo, satellite spots or nodules, spread of pigment from the edge of naevus to the normal skin.
7. *Development of symptoms:* Itching, awareness of the lesion, serous discharge, bleeding.
8. *Regional lymphadenopathy.*

Any change in a naevus as mentioned above deserves immediate attention to exclude malignant change. *Always biopsy a suspect pigmented lesion.*

Treatment of naevus

Surgical excision is indicated under the following circumstances:

1. For cosmetic reasons:
 - (a) Naevi appearing at birth – usually innocent.
 - (b) Naevi appearing before puberty – usually junctional naevi.
2. Any naevi situated in the potentially dangerous areas like soles, palms, digits, genitalia, and mucous membrane.
3. Any naevi situated in the area subjected to repeated trauma, e.g., rubbing by clothes, brassiere strap, belts, or cuts while shaving.
4. Giant hairy mole.
5. Suspicion of malignant change in a mole – see above.

Principles of excision

The operation is called **total excision biopsy**.

1. Naevi appearing at birth or before puberty – *can be excised close to their margins.*
2. All other naevi, particularly the suspicious lesion – the excision should be complete and *should include a margin of normal skin at least 2 mm beyond the margin of the lesion.*

Any excised naevus should be sent for histopathological study. If histopathology confirms the diagnosis of malignant changes with positive margin then *a wider excision is performed depending on the thickness of the tumour.*

Microscopic evidence of malignancy includes hyperchromasia, mitotic figures, pleomorphism, anaplasia and subepithelial spread.

MALIGNANT MELANOMA

This is a malignant tumour arising from the epidermal melanocytes. Melanocytes are pigment producing cells which are normally located in the basal layer of epidermis.

Malignant melanoma may develop in any area of the skin – **cutaneous melanoma**.

Malignant melanoma may also arise in the pigmented area of the eye – **ocular melanomas**, or in the mucosa – **mucosal melanoma**.

CUTANEOUS MELANOMA

This is one of the most malignant and dangerous forms of cancer of the skin. This is also a most rapidly infiltrating tumour. Fortunately this is a rare tumor accounting for only 3 percent of all malignant tumour of the skin.

Origin

1. De novo – 10 percent of cases.
2. In some pre-existing mole such as junctional naevus, compound naevus, freckles – 90 percent of cases.

Aetiology

1. *Age*: Common after puberty, usually 3rd decade i.e. usually in the 20 and 40 years of age.
2. *Sex*: Occurs most commonly in the female. Mortality is slightly higher in male.
3. *Race*: More common in fair skinned people who have been exposed to sunlight for a longer period, that is why it is more frequent in Queensland, Australia.
Rare in negroes, but survival is poor in blacks.

4. *Factors associated with increased risk:*

(a) Genetic predisposition

- Fair-skinned persons with red hair and freckles.
- Albinism
- Multiple atypical or dysplastic naevi
- Xeroderma pigmentosa
- Persons born with congenital giant melanocytic naevi
- Family history of melanoma.

(i) About one in every ten patients diagnosed with this disease has a family member with a history of melanoma.

(ii) Each person with a first degree relations diagnosed with melanoma has a 50 percent greater chance of developing this disease (cf. breast carcinoma)

(iii) FMMM (Familial Atypical Multiple Mole Melanoma) Syndrome: People with this syndrome are at greater risk of developing melanoma. Gene mutations to p53 and BRAT have been associated with familial melanoma.

(iv) Children of melanoma prone families may develop melanoma before adolescence.

(b) Environmental factors:

- Sun exposure is the only avoidable factor.
- Ultraviolet radiation (UVA & UVB) are demonstrated to be carcinogenic.

- Increased incidence at the sites exposed to sunlight. Rarely occurs on double covered areas.
- Solar irradiation is becoming much more dangerous due to the decreasing ozone layer in the atmosphere.

(c) Increased incidence of melanoma has been noted in immunosuppressive patients as in patients with renal transplantation and leukemia.

5. *Role of trauma*: Not well-defined but high incidence of melanoma in the sole has been observed in Bantu-blacks who walk barefoot.

6. *Role of pregnancy*: Pregnancy is wrongly alleged to provoke malignant change in a naevus. Though pigmentation is increased during pregnancy it has no effect on melanocytic neoplasm.

- The treatment policy should not be altered in pregnant women.

- Women who have had treatment of melanoma are advised to avoid pregnancy for 3 years.

- Often there is a tendency for malignant melanoma to regress during pregnancy.

7. *Site of predilections:*

- Cutaneous melanomas can occur in any part of the body including genitals.

• Distribution:

- 25% in head and neck
- 25% over trunk
- 25% in lower limb
- 11% in upper limb
- 14% on genital

- *In females*: Lesions are more common on the lower extremities. *In males*: Lesions are more common on the trunk and head & neck.

- *In whites* – more than 90% of melanomas occur on the sun exposed skins.

- *In blacks* – more than 60% of melanomas arise on non-sun exposed skin, involving in particular planter, subungual and mucosal surfaces. In Bantu tribe, sole is the commonest site.

- 70% of melanomas in blacks are found on the lower limb and 90% of the lesions of the leg occurs below the ankle.

- Other sites:

- Eyes (iris, ciliary body, choroids)
- Mucocutaneous junction (anorectal region, genitals)
- Head & neck (meninges, nasopharynx, paranasal sites).

8. Progress

- The course of the disease is unpredictable. As a rule there is early and widespread dissemination which leads to high mortality.
- It has been reported that spontaneous regression may occur in exceptional cases.

Malignancies which can spread from mother to foetus

- Malignant melanoma
- Lymphosarcoma

Surgical pathology

Pathologically malignant melanoma is regarded as melanocarcinoma. Rarely it may appear like a sarcoma.

Clinicopathological classification of Malignant Melanoma

Lentigo Maligna Melanoma (LMM)

- Least common and least malignant.
- Slowly growing and remains non-invasive for many months.
- Often arises in Hutchinson's melanotic freckle (HMP) consequent often on prolonged solar irradiation.
- *Most frequently seen in elderly.*
- Developed on the skin areas with high levels of sun exposure e.g., in the face or arms.
- A superficial melanoma confined to the topmost layers of the skin is *often presented as a brown or black stain on the skin (macular).*
- *Infiltrated nodule develops in an irregularly pigmented macule which has been present for several years.*
- Lymph node metastases are rare and occur later in clinical history.

Superficial Spreading Melanoma (SSM) (Fig. 2.58A)

- Most common variety but less aggressive lesion.
- Occurs on any part of the body but commonly lower limbs in women and trunk in man.

- Commonly arises on a pre-existing naevus.
- Age group is younger than in LMM.
- The lesion is usually present as a *flat or elevated plaque (papular) with irregular border and spreading orientation on the skin with colour variation* – brown, black, blue or pink.
- *The lesion is usually palpable. Nodule may develop within the lesion; there may be ulceration.*
- Intermittent itch is a common symptom.
- *Lateral or radial growth predominates.*
- Lymph node metastases is rare until a nodular component develops.

Nodular Melanoma (NM) (Fig. 2.58B)

- Least common but most aggressive.
- It can occur on any part of the body.
- *Nearly all mucosal, genital and anal melanomas are nodular ab-initio.*
- *Presents as a well-demarcated smooth nodule or lump (nodular) and well-pigmented.* It appears in a mole or on normal skin.
- It rapidly grows larger. *Vertical growth is the prominent features in contrast to the SSM where lateral growth predominates.*
- Frequently exophytic, i.e., it demonstrates upward growth.
- *Itch is common and ulceration occurs more frequently leading to weeping or bleeding.*

Acral Lentiginous Melanoma (ALM) (Figs. 2.58C, D, E)

- Most aggressive lesion with early metastasis.
- Such lesions are more common in oriental and black persons.
- *Occurs mainly in the skin of palm and sole, and under the nail (subungual melanoma)*
- A variety of SSM but occurs on sites that are protected from sun burn by a thick layer of keratin.
- **Special varieties of ALM**
 - (a) *Plantar melanoma:* Common in blacks. Usually heavily pigmented. Ulceration and fungation occur in advanced lesions.
 - (b) *Subungual melanoma:*
 - Usually begins as a pigmented linear streak in the nail bed.
 - Symptom less in early stage. Often attention is drawn to the lesion only after injury.
 - Distal migration of pigment away from the

MELANOMA (Contd.)

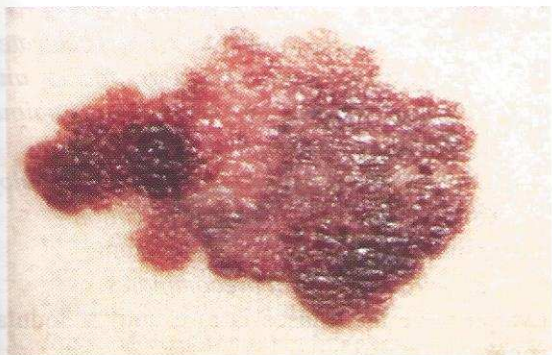


Fig. 2.58A. Superficial spreading melanoma. Note the irregular margin of the lesion with spreading orientation, colour variations from brown to black, and irregularity of the surface.



Fig. 2.58D. Malignant melanoma right heel.



Fig. 2.58B. Nodular melanoma. Note the well demarcated smooth reddish lump arising on a pigmented mole.

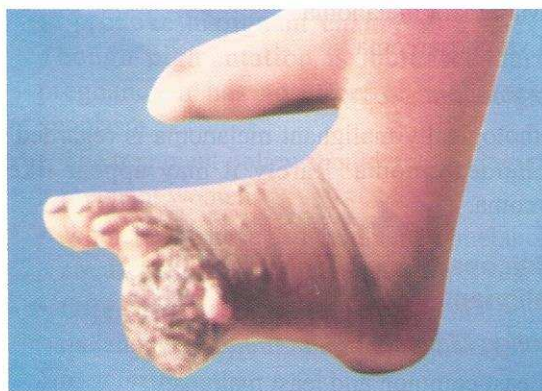


Fig. 2.58E. Malignant melanoma involving the little toe. Note also the subcutaneous nodules (satellite nodules) on the dorsum of the foot.

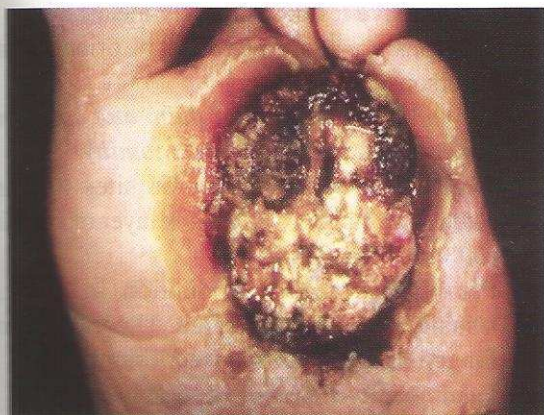


Fig. 2.58C. Acral lentiginous melanoma. Note the pigmented proliferative lesion on the sole of the left forefoot. Wedge biopsy suggested malignant melanoma. Conservative below knee amputation was performed. Patient was sent to oncologist for further therapy.

nail bed, pigmentation of nail fold (Hutchinson's sign) and progressive elevation of the nail from the nail bed will lead to suspicion of subungual melanoma. In late stage, the tumour may extrude through the nail plate when ulceration, secondary infection and bleeding are common.

D/D of subungual melanoma: Subungual melanoma should be differentiated from the following lesions:

- Subungual haematoma
- Pyogenic granuloma
- Chronic paronychia
- Bacterial and fungal infections
- Rare vascular tumour

Histological classification

- Depends on the course and thus anatomical level of invasion to determine the microstage of the melanoma.
- Very useful in predicating the type of treatment and the prognosis.

Course

Most malignant melanoma grow in 2 phases:

- *Phase of radial growth (superficial growth)*: Lesion extends superficially and laterally. The neoplastic cells are confined to the epidermis.
- *Phase of vertical growth*: Malignant cells invade the dermis and deeper tissues.

Vertical growth has a worse prognostic variable than radial growth.

Vertical growth can be evaluated by two ways:

- Clark's anatomic level of invasion.
- Breslow's tumour depth measurements.

Both the "Clark level" and "Breslow's tumour depth" refer to the microscopic depth of tumour invasion.

Clark's anatomic level of invasion

- Level I • Tumour cells confined to the epidermis above basement membrane – **melanoma-in-situ**.
- Do not develop metastasis i.e. non-invasive
- Level II • Tumour cells invade the papillary dermis after having broken through the basement membrane.
- Rarely produce metastasis.
- Level III • Tumour cells extend to the interface between papillary and reticular dermis. A portion may protrude, but does not invade the reticular dermis.
- A small but significant proportion will develop metastasis.
- Level IV • Tumour cells extend into the reticular dermis.
- Level V • Tumour cells extend into the subcutaneous tissue.
- Both levels IV and V: A large proportion will develop metastasis.
- Nodular melanomas are regarded as being at least level III in depth of invasion.

Breslow's Tumour Depth (Thickness) measurement

- *Measured the vertical thickness of tumour in millimeters*: Breslow introduced staging by measuring the maximal vertical thickness of the melanoma from the top of the granular layer of the epidermis to the deepest melanoma cells in the dermis.
 - Breslow's method of measurement requires an optical micrometer fitted to the ocular position of a standard microscope.
- Stage I Thickness 0.75 mm or less
- Stage II Thickness 0.76 mm to 1.50 mm
i.e. < 1.51 mm
- Stage III Thickness 1.51 mm to 3.00 mm
i.e. ≤ 3.00 mm
- Stage IV Thickness more than 3.00 mm
- Breslow's tumour depth measurement is reliable and more accurate as a prognostic index and thus is important determinant of therapy.

A comparison of Clark's level of invasion and Breslow's tumour depth can be assessed to predict the virulence of the lesion (Fig. 2.59).

Clark's level of invasion	Breslow's tumor depth	Virulence
Level I	0.75 mm from the surface of the skin (epidermal lesion)	Low risk primary tumour. Do not metastasize at any site
Level II } Level III }	0.76 mm to 1.50 mm	Intermediate risk. 25% incidence of metastasis.
Level IV } Level V }	1.51 mm deeper below the skin surface	High risk. 60% incidence of metastasis.

Spread

Malignant melanoma may spread by local extension, by lymphatics or by blood stream.

Local spread

- Initially, the growth spreads *laterally* by continuity and contiguity within the epidermis i.e., intraepidermal growth – *radial growth phase*. Metastasis is unusual.
- Later the growth spreads *vertically* within the dermis – *vertical growth phase*.

- covered with a crust of blood and serum. Bleeding is common on the surface. A big melanoma may look wet, soft and boggy from infection and bleeding.
13. *Size and shape*: Varies, the border is usually irregular. A neglected nodular melanoma will become a large florid tumour which protrudes from and overlaps the surrounding skin.
 14. *Consistence*: The primary tumour feels firm and solid. The satellite nodules feel hard.
 15. *Temperature and tenderness*: Temperature remains the same as that of the surrounding skin. *The tumour is also not tender.*
 16. *Mobility*: The tumour is intimately fixed to the skin and moves with the skin. It can be easily lifted up from the deeper structures.
 17. *Surrounding skin*: There may be *inflammatory halo* or a *halo of brown pigment* in the skin around the tumour. *Satellite nodules*, pigmented or depigmented, *intransit metastasis*, may be seen or felt in the skin and subcutaneous tissues between the primary tumour and the nearest draining lymph nodes.
 18. *Regional lymph nodes*: Should be examined. They may be enlarged.

General examination

19. One should examine for distant metastases, e.g., liver (hepatomegaly, jaundice), lungs (pleural effusion), brain (neurological abnormalities), bones (pathological swelling or fracture).

Five important features of melanoma suspicion in a pigmented lesion: ABCDE

- Asymmetry
- Border irregularity
- Colour variation
- Diameter > 6 mm
- Elevation

Clinical staging

Clinical staging depends on the presence or absence of satellite nodules, intransit metastasis, clinically involved lymph nodes and/or distant metastases.

- Stage O: Melanoma-in-situ. The melanoma cells are found only in the outer layer of the skin and have not invaded the deeper tissues i.e., *intraepidermal growth*.

No nodal involvement. Thus localised to the site of origin (cf. Clark Level I).

- Stage I: Thin melanoma (≤ 1.50 mm)
Rarely nodal involvement (cf. Clark Levels II & III).
- Stage II A: Thick melanoma between 1.51 to 4 mm (cf. Clark Level IV).
- Stage II B: Thick melanoma more than 4.00 mm. or penetration into the subcutaneous tissue (cf. Clark Level V) or 2 mm thick melanoma with surface ulceration.
- Stage III: Any tumour size + spread beyond the primary site of origin i.e. presence of *sattelosis*, *intransit metastasis*, *regional lymph node metastasis* or all of them.
- Stage IV: Any of the above + distant metastasis such as involvement of two or more groups of lymph nodes; visceral metastases e.g. lung, liver, bone and brain are the more frequent sites; disseminated cutaneous metastasis.

Treatment

1. Treatment of the primary lesion.
2. Treatment of the regional lymph nodes.

Treatment of the primary lesion

- Surgical excision of the primary lesion after confirmation of diagnosis by adequate biopsy is the treatment of choice.
- Radiotherapy and chemotherapy have no role in the management of primary melanoma.

Establishment of diagnosis

- *Excision biopsy of a suspected lesion is only recommended* to confirm diagnosis and provide histopathological information that guides further surgical strategy.
- A tridimensional excision biopsy should be performed *with a margin of at least 2 mm lateral clearance beyond the extent of the lesion*.
- The orientation of incision should be elliptical in shape in the direction of langer's line to allow tension free closure.
- The excision must include underlying cuff of subcutaneous tissue deep to the tumour upto the deep fascia so that complete histological data

including Clark's levels of invasion and Breslow's tumour depth can be assessed.

- *The deep fascia should not be interfered with or dissected* to avoid spread of melanoma to the tissues deep to the deep fascia.
- *Excision biopsy on the extremities* should be in the longitudinal direction of the limb to allow for a primary wound closure.
- *Use of wide local excision (WLE) as an initial biopsy* may reduce the efficacy of subsequent lymphatic mapping required to know the lymphatic spread.
- *Incision and needle biopsy should be avoided in melanoma* because of the risk of transferring melanoma cells to the subcutaneous fat and deeper tissues and thus conversion of a superficial melanoma to a deep melanoma.
- *A punch biopsy or excision of a segment of the lesion* may be appropriate if complete excision causes unreasonable deformity e.g., in large lesions of the face for cosmetic reason.
- *Shave biopsy or curettage should not be used* when melanoma is suspected because the cancer cannot be staged accordingly.
- For any suspicious or persisting lesions of the nail, an adequate biopsy material is required for histological examination and should include a portion of the proximal terminal of the nail matrix, the nail bed and the nail plate.

Once the diagnosis of melanoma is confirmed, investigations to look for lymphatic spread and metastasis are carried out.

- Investigations for lymphatic spread
 - In presence of enlarged regional lymph node(s)*: FNAC of the enlarged node(s) is helpful to detect the lymphatic spread and to stage the disease.
 - In absence of enlarged lymph nodes*: Sentinel lymph node biopsy (SLNB) following lymphatic mapping is required to detect the lymphatic spread.
- Investigations for metastasis
 - Chest X-ray
 - LFT
 - LDH
 - Pre-operative liver, brain and bone scan:
 - Not mandatory, for clinical stage I lesion.
 - Should be done if LFT, LDH is abnormal.

- Extraordinarily high LDH often indicate metastatic spread to the liver.

Tumour markers for melanoma:

- Melan-A
- S-100
- HMB45 (Hydroxy Methyl Bromide)
- LDH

Treatment of the primary melanoma

- *Surgical resection of primary melanoma is the treatment of choice*
- After histological diagnosis, the site of the original lesion is usually reexcised with adequate margins dictated by the tumour thickness. Often this is done by a *wide local excision (WLE)* with 0.5 to 3 cm. margins. *This wide excision aims to reduce the rate of tumour recurrence at the site of the original lesion.*

Recommended margins for surgical resection of primary melanoma

Primary melanoma	Margin radius (cm)
(i) Melanoma in situ	0.5 cm
(ii) < 1.0 mm	
(iii) = 1.0 mm	1.0 cm
(iv) > 1–2 mm	2.0 cm
(v) > 2 mm	3.0 cm

- The 3 cm margin of excision is also advocated for tumours with regression noted on histological examination.
- For lesion in the face, maximum safe margin of excision is 2.5 cm for cosmetic reason.
- The deep fascia is not included in the excision because it is noted that the deep fascia limits the local spread or 'in-transit' recurrence to the subcutaneous tissue.
- The residual defect following WLE is preferably closed directly by primary suture. Where direct closure is not technically possible a split-skin graft can be used. Closure developed by rotational or advancement flaps or pedicle flap is better avoided because a thick flap may hide a local recurrence for longer period of time.

SUBUNGUAL MELANOMA

Amputation at the metatarsophalangeal or metacarpo-

phalangeal joint is the optimal treatment. Cade advised removal of the metatarsal or metacarpal also (i.e., Ray amputation)

MELANOMA OF THE SOLE OF THE FOOT OR PLANTAR MELANOMA

- (i) *Amputation is seldom performed now-a-days*, because of improvements in non-ablative therapy available in certain centers.
- (ii) *Wide excision of the lesion with a large surface area is combined with isolated limb perfusion with Melphalan*. This combined therapy is found to be effective in patient with planter and subungual melanoma with minimal morbidity.
The residual large defect may present a difficult problem during reconstruction. *A split skin grafting generally provides effective cover* and patients are able to walk on the grafted foot with little difficulty after using a special foot wear.
- (ii) Limited amputation of the limb may be justified in advanced case to remove a foul smelling fungating mass.

Treatment of regional lymph nodes

- Prophylactic lymph node dissection (PLND)
- Elective lymph node dissection (ELND)
- PLND = ELND
- Therapeutic lymph node dissection (TLND)
- Sentinel lymph node biopsy (SLNB)
- The role of prophylactic lymph node dissection (PLND) is controversial and avoided.
- Melanomas which spread usually do so initially to the regional lymph nodes before spreading elsewhere.
- Attempts to improve survival by regional lymphadenectomy are associated with many complications but unfortunately no survival benefit. **So, PLND is not advocated now-a-days.**
- In absence of clinical regional lymphadenopathy, recently the technique of sentinel lymph node biopsy (SLNB) is being performed to note the presence of lymph node metastases to justify the need of lymphadenectomy and thus to reduce the complications of lymphadenectomy.
- **SLNB is advocated in following conditions:**
 - (i) Primary tumour thickness ≤ 1 mm but with surface ulceration

(ii) Primary tumour thickness > 1 to 2 mm.

(iii) Primary tumour of the limb.

- SLNB is being tried both in impalpable or palpable nodes.
- **Alternatively in presence of clinically palpable node/nodes, FNAC is performed.**
- Routine H&E staining and immunoperoxidase staining will be adequate to rule out nodal involvement. Step dissection & PCR tests on nodes may be performed also to detect micrometastases.
- **In presence of lymph node involvement as detected by SLNB or FNAC – TLND is performed.**
- TLND is also advocated, if during follow up previously normal lymph nodes becomes enlarged and suspicious.
- *TLND should be avoided in patients with primary lesions greater than 4 mm in thickness* because of the high risk of blood borne metastasis at the time of initial diagnosis.
- TLND is also avoided in patients with stage IV diseases with distant metastases, or with advanced age and/or poor physical state as the long term survival is poor.
- *Where inguinal nodes are involved*, radical ilio-inguinal dissection (used to be performed before) is not justified because long term survival after removal of involved iliac nodes is extremely poor.

Monoblock dissection

During TLND, where possible, i.e., on the trunk, on the limbs above the elbow or knee joints, on the face, the primary tumour is excised along with block dissection of the regional nodes *in continuity* (monoblock dissection). Monoblock dissection removes all the potentially contaminated tissues between the primary lesion and the draining lymph nodes and thus prevents in-transit recurrence.

For facial primary lesion – superficial parotidectomy is also advised.

Complications of block dissection

- Particularly when concerned with the inguinal nodes – block dissection is associated with considerable morbidity, such as necrosis of skin flaps, wound dehiscence, wound infection, lymph fistula, life long oedema or swelling of the limb.

- In the upper limb the same morbidity is less encountered.
- After radical inguinal node dissection, transposition of the sartorius muscle is done to cover the femoral vessels.
- Once the lymph nodes dissection is completed the patient will be considered for adjuvant therapy.

Sentinel lymph node biopsy (SLNB)

- First described by CABANA in 1977.
- Sentinel lymph node(s) is *the first node in the lymph node basin which will receive the cancer cells first from the primary site.*
- Biopsy of the SLN(s) helps in assessing the involvement of the lymph node(s) by the cancer cells. If the SLN(s) contains cancer cells (*lymph node status is positive*), it is likely that the rest of the lymph nodes basin also contain cancer cells, so the rest of the lymph nodes are to be removed surgically.
- If the SLN(s) does not contain any cancer cells (*lymph node status is negative*), so lymphadenectomy can be spared.
- SNL(s) biopsy is done after the biopsy of the melanoma *but before the wide local excision (WLE) of tumour, as it prevents SLN mapping.*
- *Importance of SLNB:* The SLNB concept helps in staging procedure whether stage III or not.

SLN Mapping

The procedure to identify the sentinel node (s) is called sentinel lymph node mapping.

- (i) A radioactive substance ^{99m}Tc labeled in the sulphur colloid is injected intradermally around the peritumoral skin 1–4 hours before surgery. The SLN is identified by a hand held gamma camera.
- (ii) Isosulphan blue may also be injected in the tumour or peritumoral skin on the day immediately before surgery. The dye will stain the SLN(s) blue which can be identified at operation.
- (iii) The combination of radio colloid and isosulphan blue dye injections raises the accuracy of SLN(s) identification to 90%
- (iv) The surgeon then removes the sentinel node(s) for histopathology to check for cancer cells.

NB: SLNB is practised in the following malignant lesions:

1. Malignant melanoma
2. Carcinoma penis
3. Carcinoma breast

Treatment of metastatic malignant melanoma

Metastatic malignant melanoma is refractory to most modalities of treatment. In general, this tumour is *highly radio-resistant* and *relatively chemoresistant*. Immunotherapy remains still in the experimental stage. All these modalities are still considered palliative at the present time.

Radiotherapy

- **Malignant melanoma is regarded as highly radio-resistant.**
- It may reduce the rate of local recurrence but does not prolong survival.
- *So, no role in the treatment of primary tumour.*
- *Beneficial only in secondaries in brain and bone.*
- Metastatic melanomatous masses in the soft tissues have been satisfactorily treated using 800 rads $\times$ 4 weekly intervals in some European centers.
- In other centers, radiotherapy is usually reserved for the treatment of bone and cerebral secondaries; substantial palliation and pain relief can be achieved temporarily.
- **Endolymphatic therapy** – with radioactive ^{131}I or ^{32}P . Recently Endolymphatic therapy is used in many centers as a preliminary to block dissection. Radioactive ^{131}I or ^{32}P is injected with ultra fluid lipiodol as a vehicle into the lymphatic channel: the block dissection is performed about 4 weeks after the intralymphatic injection of radioactive substance.

Chemotherapy

- **Malignant melanoma is also considered relatively chemoresistant.**
- However, systemic chemotherapy has been widely used in the treatment of metastatic melanoma both with single and multiple agents.
- Drugs most commonly used are DTIC (decabazine), BCNU, Vincristine, Vindesine, Melphalan.

- Vindesine has very few side effects in comparison to DTIC and is therefore used as a first line of treatment.

Isolated limb chemoperfusion

- For melanoma of the arm or leg: chemotherapy drugs can be put directly into the blood stream supplying that limb.
- *It is designed to provide a high local concentration of cytotoxic drugs and thus causing minimal systemic toxicity.*
- This technique involves clinical isolation of limb vessels using a tourniquet, followed by cannulation of the major artery and vein supplying the limb and then introducing the cytotoxic drugs with the help of extracorporeal circulation through an oxygenated circuit in addition to a heat exchanger. This will help hyperthermic perfusion of the isolated limb with well-oxygenated blood and delivering a high concentration of cytotoxic drugs into the limb.
- Drugs commonly used Melphalan or DTIC.

Indications

- (i) Locally recurrent melanoma of the limb with multiple skin deposits (in-transit metastases).
- (ii) Prophylactic treatment of poor prognostic limb tumour e.g., tumors thicker than 1.7 mm with no evidence of nodal disease as an adjunct to surgical therapy of primary lesion.
- (iii) Clinically Stage II & III diseases.
- Regional chemo-perfusion is however associated with complications like arterial thrombosis, persistent oedema of the limb, infection, hepatitis and myelosuppression.

Immunotherapy

- There is evidence that melanoma is associated with immune responses in the host and may regress on its own, primary tumour more commonly than the metastatic deposits. Various trials have been developed to exploit this phenomenon. As yet such therapy is still in experimental stage.
- Interferon alpha and Interleukin 2 (also called IL2 or aldesleukin) are commonly used.
- Some superficial melanomas (lentigo maligna) have resolved with topical application of Imiqu-

imod (Aldara) cream, an immune enhancing agent. Some dermatosurgeons use this cream prior to surgery for melanoma-in-situ lesions located in cosmetically sensitive regions e.g. face. Some surgeons excise the melanoma surgically and then treating the area with Aldara cream postoperatively for 3 months.

Monoclonal antibodies IgG3

These are directed against the antigens expressed on the surface of melanoma cells. IgG3 activates the immune system.

Hormone treatment

Antiestrogen like tamoxifen have been tried in presence of systemic diseases with a response rate of 15–20%.

Prognosis

- The prognosis varies with the pathological type of the lesion, level of invasion, tumour thickness, clinical staging, site and sex.

5-year survival predictions

- *Low risk primary tumour:* 95–100% chance of a 5-years cure with conservative surgical treatment.
- *Intermediate risk tumour:* 85–95% chance of a 5-years cure.
- *High risk tumour:* 15–50% chance of a 5-years cure.
- *If lymph node metastasis is present* the prognosis becomes worse.
- *Surface ulceration of the lesion* lowers the survival rates for tumours of comparable thickness.
- *Melanoma of the face* has a better prognosis.
- *Subungual melanoma* has a relatively good prognosis.
- *Women overall* have a 10% survival advantage.

NON-CUTANEOUS MELANOMA

- Mucosal melanoma
- Ocular melanoma.
- Rarely, melanoma may be found in the intestines and in the meninges.

Mucosal melanoma

- Melanoma may occur in the mucosa also.
- Mucosal melanoma are common in blacks.

- Involves the mucosa of the lips, nose, palate, genital tract and anorectal region.
- Irrespective of the site, mucosal melanomas are associated with a poor prognosis owing to early spread to lymph nodes and to the advanced stage of the lesion at diagnosis.
- Anorectal melanoma is treated by abdominoperineal resection.
- Melanoma vulva is treated by vulvectomy with plastic reconstruction.

Ocular melanoma

- Accounts for 2% to 5% of all melanomas.
- May occur in the eye at the following sites:
 - (i) *the uveal tract* which comprises the choroids, ciliary body and iris.
 - (ii) *the conjunctiva*.
- Often diagnosed as reduction of vision, a vitreous haemorrhage, or by the chance finding of an elevated pigmented lesion on the conjunctiva.
- More posterior the lesion the more malignant it is likely to be.
- Displays distinct pattern of behaviour:
 - (i) No lymphatic drainage, so no nodal metastasis.
 - (ii) Hepatic metastasis – most common site of metastases.
- Spread is often delayed for many years and so associated with the longest survival from the diagnosis of primary tumour to recurrences.
- A history of enucleation of an eye with or without replacement by a glass eye due to a disease (not known) long ago and an appearance of an enlarged liver recently should suggest an ocular melanoma for the lost eye.

Treatment:

- (i) Malignant melanoma of uvea:
 - Laser photocoagulation – small melanomas of the choroids and > 2 mm away from the macula and optic disc.
 - Plaque brachytherapy – medium-sized choroidal and ciliary body melanomas.
 - Resection – *local resection* is usually reserved for small lesion of the iris, ciliary body and choroidal melanomas.
 - *Enucleation* is done when the tumour is too large to manage with radiotherapy or there

is invasion of optic disc or there is no chance of visual recovery.

- *Exenteration of the orbit* – It is considered when there is extensive extraocular involvement without any metastasis or in presence of orbital recurrence after enucleation.

(ii) Conjunctival melanoma:

- Complete surgical excision using no-touch technique.
- Adjuvant cryotherapy to the edges of the conjunctiva after excision.
- Topical chemotherapy with Mitomycin C after complete healing of the ocular surface.

Pigmented lesions of the skin other than melanoma

1. Seborrhoeic keratoses
2. Pigmented basal cell carcinoma or squamous cell carcinoma
3. Cutaneous angiomas: The soft consistency of the lesion and blanching on pressure are often helpful in making the diagnosis.
4. Pyogenic granuloma
5. Subungual haematoma
6. Glomus tumour
7. Kaposi's sarcoma
8. Histiocytoma
9. Keratoacanthoma
10. Epidermal naevus

When doubt exists in the diagnosis, excision biopsy is indicated in the above mentioned lesions to exclude malignant melanoma.

11. Cafe-au-lait patch: This is an area of pale brown milk-coffee coloured pigmentation in the skin. This is often associated with neurofibromatosis and sometimes with pheochromocytoma. This is present at birth and never undergoes malignant change.
12. Multiple circumoral melanosis associated with the Peutz-Jeghers syndrome. The Peutz-Jeghers syndrome is a familial condition and consists of:
 - multiple intestinal polyposis, and
 - multiple melanin spots around the mouth, on the lips and in the buccal mucous membrane. These moles never turn malignant.

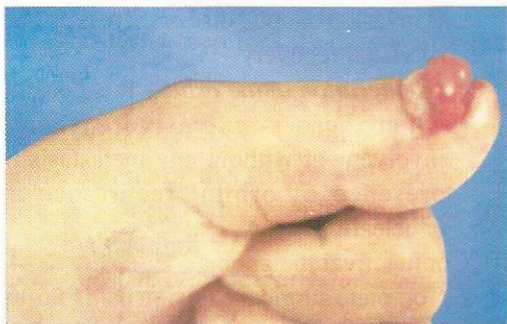


Fig. 2.60A. Pyogenic granuloma at the nail bed of the left thumb. D/D: Haemangioma, malignant melanoma.



Fig. 2.60B. Pyogenic granuloma of left little finger.

PYOGENIC GRANULOMA

It is an exuberant overgrowth of granulation tissue producing a red, soft or moderately firm, polypoid lesion typically 5 to 10 mm across. The lesion looks like a haemangioma but has a typical natural history. It develops due to abnormal inflammatory reaction

in response to trauma or a foreign body, such as a splinter. The history of trauma may not be remembered by the patient. Resolution is very slow. Sometimes ulceration develops when differential diagnosis from the malignant lesion becomes difficult.

Treatment is excision.

3

CHAPTER



Swellings in the Neck

Swellings in the neck are caused by innumerable pathological lesions arising from the anatomical structures lying therein.

Skin and superficial fascia

- Sebaceous cyst
- Lipoma
- Fibroma

Lymphatics

- Cystic hygroma
- Solitary lymphatic cyst

Lymph nodes

- Inflammatory
- Neoplastic
- Reticuloses

Blood vessels

- Aneurysm
- Haemangioma
- Carotid body tumour

Nerve tissue

- Neurofibroma

Thyroid gland

- Inflammatory
- Neoplastic
- Autoimmune disease

Pharynx

- Pharyngeal pouch

Larynx

- Laryngocele

Branchial arch remnant

- Branchial cyst

Thyroglossal duct remnant

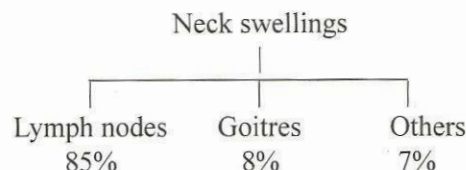
- Thyroglossal cyst

Salivary glands

- Inflammatory
- Neoplastic
- Autoimmune disease

Most of the neck swellings can often be diagnosed from the history and physical signs alone.

The most common swellings in the neck are of lymph node origin and the most likely causes of lymphadenopathy are infections and metastatic spread of cancer.



Lymph nodes are recognised by their physical signs more easily when multiple than when solitary.

Solitary lymph node swelling presents the greatest diagnostic challenges.

Goitres are swelling of the thyroid gland. They are distinguished from other lump in the neck by the fact that they move on swallowing.

Others are solitary swellings arising from variable structures.

Any solitary swelling in the neck must be distinguished from an isolated enlarged lymph node.

SURGICAL ANATOMY

Triangles of the neck (Fig. 3.1)

The anterolateral part of the neck is conventionally divided into an anterior and a posterior triangles by the diagonally running sternomastoid muscle. By pressing the jaw laterally against resistance of one's hand the opposite sternomastoid becomes taut. This muscle helps to define two triangles.

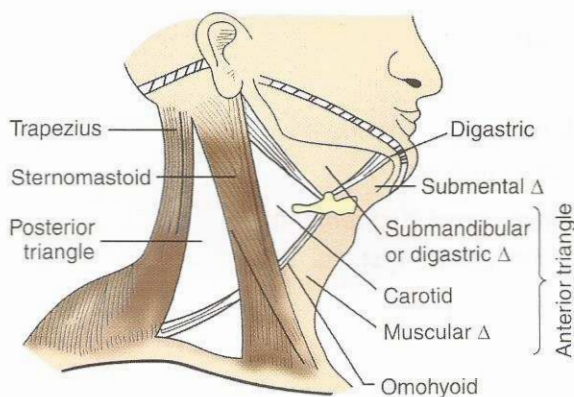


Fig. 3.1. Triangles of the neck. Some swelling is peculiar to a particular triangle, e.g., cystic hygroma in the posterior triangle.

The *anterior triangle* is bounded by the midline from chin to manubrium, posteriorly by the anterior border of the sternomastoid and above by the lower border of the mandible. It can be subdivided by the digastric muscle and the omohyoid muscle into the *suprahyoid* – *submental* and *submandibular triangles*, and the *infrahyoid* – *carotid* and *muscular triangles*.

The *posterior triangle* is bounded anteriorly by the posterior border of the sternomastoid, posteriorly by the anterior border of the trapezius and below by the middle third of the clavicle.

For clinical evaluation, the swellings in the neck

can be grouped in one of these easily definable triangles.

CLASSIFICATION

The swellings of the neck can be classified in two groups: those situated in the midline and within approximately 2 cm on either side of the midline (*midline swellings*), and those situated lateral to this area (*lateral swellings*).

Midline swellings – Cystic and solid (Fig. 3.2)

Cystic swellings

1. Ranula
2. Cervical dermoid
3. Subhyoid bursal cyst
4. Thyroglossal cyst
5. Cyst in relation to the isthmus of the thyroid gland
6. Cold abscess in the space of burns
7. Aneurysm of the innominate artery.

Solid swellings

1. Lymph node swelling:

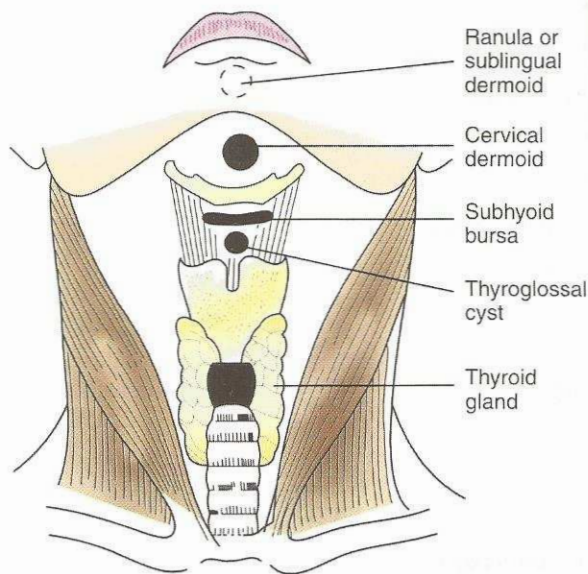


Fig. 3.2. Midline swellings of the neck.

- Submental
 - Pre-laryngeal
 - Pre-tracheal
2. Thyroid gland swelling (*moves up with swallowing*):
 - Diffuse swelling

- Solitary nodule
 - Multinodular
3. Bony growth arising from the manubrium sterni
 4. Rarely persistent thymus
 5. Rarely ectopic thyroid

All the midline swellings are included within the anterior triangle.

Lateral swellings – Cystic and solid (Fig. 3.3)

For better clinical assessment lateral swellings can be grouped according to their locations within any of the three triangles of the neck:

- (a) sub-mandibular triangle;
- (b) carotid triangle; and
- (c) posterior triangle.

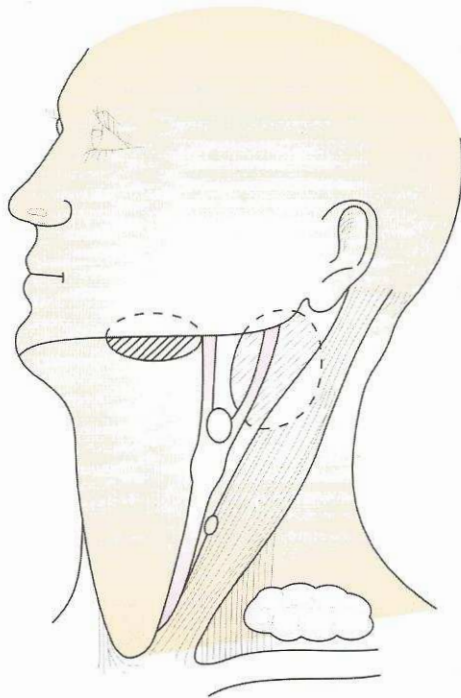


Fig. 3.3. Lateral swellings of the neck.

Submandibular triangle

Cystic swellings

1. Plunging ranula
2. Lateral variety of sublingual dermoid
3. Retention cyst of salivary gland

Solid swellings

1. Submandibular salivary gland

- Tumours
 - Sialitis
 - Sialolithiasis
 - Sjogren's syndrome
2. Lymph node swelling

Carotid triangle

Cystic swellings

1. Branchial cyst
2. Abscess in the lymph glands, e.g., cold abscess
3. Carotid aneurysm
4. Cystadenoma of lateral lobe of thyroid
5. Laryngocele

Solid swellings

1. Carotid body tumour
2. Lymph node swelling
3. Solid swelling of lateral lobe of thyroid
4. Sternomastoid tumour
5. Branchiogenic carcinoma

Posterior triangle

Cystic swellings

1. Cystic hygroma
2. Solitary lymphatic cyst
3. Abscess in the lymph nodes, e.g., cold abscess
4. Pharyngeal pouch
5. Subclavian aneurysm

Solid swellings

1. Lymph node swelling
2. Cervical rib

Swellings anywhere in the neck

In addition to the above mentioned lesions the following lesions can occur anywhere in the neck. These are:

1. Sebaceous cyst
2. Lipoma
3. Fibroma
4. Neurofibroma
5. Haemangioma.

RANULA

Ranula is a soft and bluish swelling simulating frog's belly. The term '*ranula*' is derived from the Latin word, *Rana*, which means a little frog.

Origin

It is a *mucus-retention cyst* arising from the mucous glands situated on the floor of the mouth and the undersurface of the tongue. The names of the glands are glands of Blandin and Nuhn. The swelling is mostly, if not entirely, unilateral.

According to some, it is a dilatation of the duct of the submandibular gland though this theory is not universally accepted (W. Boyd).

Pathology

Lining of the cyst wall: The wall of the cyst is lined by either *columnar or cuboidal epithelium* or by fibrous tissue.

Contents: Viscid and glairy or jelly-like fluid.

Diagnostic criteria

Once seen, diagnosis is never to be in doubt. It is obvious from its site and appearance.

1. **Age:** Mostly seen in young children and adolescence.
2. **Sex:** Both the sexes are equally affected.
3. Patient presents with *swelling in the floor of the mouth* which may be painful.
4. Intrabuccal swelling situated in the floor of the mouth between the undersurface of the tongue and the symphysis menti (Fig. 3.4), *mostly unilateral, on one side of the frenulum linguae* (patient should be asked to show the under-surface of the tongue).

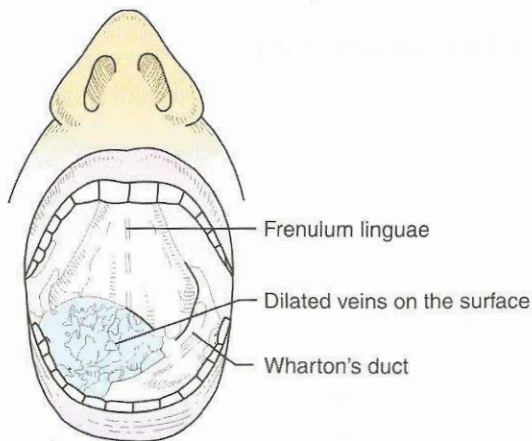


Fig. 3.4. Ranula. A typical ranula passing submucosally across the floor of the mouth with dilated veins on the surface which gives the appearance resembling a frog's belly.

5. Shape and size: Spherical cyst varies from 1 to 5 cm in diameter but only the top half is visible.
6. *Colour is pale blue with characteristic semi-transparent appearance* and with the presence of ranine vein (deep lingual vein) found as a bluish line over the surface.
7. Surface is smooth and mucous membrane is mobile over the swelling. The edge of the swelling is difficult to feel. The swelling is not tender.
8. *Cystic swelling: Fluctuation is positive.* (Fluctuation is elicited with the tip of the index finger, and is called *Paget's test*, as a cystic swelling feels more cystic in the centre.)
9. *Transillumination: Positive.* During transillumination amidst the reddish glow there is the presence of white opaque strands which indicate either the presence of deep lingual veins running along the wall of the cyst, or Wharton's duct.
10. Patient may give the history of bursting of the ranula with subsequent reformation.
11. May or may not have prolongation in the neck.
12. Cervical lymph glands are usually not enlarged.

Types of ranula

Oral type (Fig. 3.4): Simple ranula restricted to the oral cavity (features already described).

Plunging type: When the oral variety extends into the neck (particularly submandibular region) passing beyond the floor of the mouth along the posterior border of mylohyoid muscle and appears in the submandibular region.



Fig. 3.5. Ranula on the floor of the mouth on the right side of frenulum linguae.

Diagnosis of plunging ranula: By bidigital palpation of the swelling. One finger is placed in the oral cavity and the other on the neck. If the pressure is given from the neck, finger in the oral cavity is pushed up, and vice versa.

Therefore, in all cases of ranula, one should examine the submandibular region for any cervical prolongation.

Complications

1. Repeated trauma
2. Bursting and reformation
3. Infection
4. A big ranula may cause difficulty in speech or eating.

Differential diagnosis

1. Sublingual dermoid: Midline swelling; transillumination is negative.
2. Haemangioma: Compressible; transillumination negative.

Treatment

1. Complete excision.
2. Partial excision with "marsupialization".

Complete excision is ideal, but it is difficult and tedious, because it almost always bursts on attempted dissection, and there is a chance of incomplete removal and so, a chance of recurrence.

Partial excision with marsupialization: The top of the cyst is excised. The cut edge of the cyst wall is sutured with the mucous membrane covering the floor of the mouth. Thus the bottom of the ranula becomes part of the floor of the mouth.

Every care should be taken not to injure the Wharton's duct.

Plunging type ranula is better approached through the neck along the Langer's line over the submandibular region.

CERVICAL AND SUBLINGUAL DERMOID

This is a form of congenital sequestration dermoid arising due to inclusion of the surface ectoderm at the fusion line of the 1st branchial arches or mandibular arches.

Common age of appearance

Though congenital, it does not appear before the age of ten. The usual time of appearance is between 10 and 25 years.

Pathology

Sublingual dermoid is a thin-walled cystic lesion.

Lining of the cyst wall: As it is a *congenital ectodermal* swelling, it is lined by squamous epithelium.

Contents: Cheesy material or sebaceous material. It never contains hair as in other dermoids.

Types

According to position:

1. Median variety
2. Lateral variety (uncommon)

Each variety is again either above the mylohyoid muscle or below the mylohyoid muscle. So, anatomically it is of two varieties:

1. Supramylohyoid or sublingual
2. Inframyllohyoid or cervical.

Pathogenesis

During fusion of the two halves of the mandible some ectodermal cells may be sequestered into the underlying mesoderm. In future these ectodermal cells may proliferate and ultimately liquefy to form the cystic swelling. The swelling may be projected below the mylohyoid muscle (inframyllohyoid variety), or above the mylohyoid muscle (supramyllohyoid variety) in the floor of the mouth below the undersurface of the tongue and hence is called sublingual dermoid.

According to some, the cervical type of dermoid is derived from the 2nd branchial cleft, particularly the lateral variety.

Diagnostic criteria

The patient may present with the swelling in different manners according to the positions.

1. Median variety

(a) Supramyllohyoid variety or sublingual variety

1. *Midline cystic swelling in the floor of the mouth between the tongue and the jaw.*



Fig. 3.6. Sublingual dermoid. Patient having this swelling in the floor of the mouth beneath the tongue. Duration – for the last 20 years. Fluctuant, non-opaque swelling. Patient did not have difficulty in eating but she had some difficulty in speech.

2. Shape and size: Spherical cyst varies from 2 to 5 cm in diameter.
3. Surface is smooth. The swelling is not tender.
4. Cystic swelling – *fluctuation is positive*.
5. Transillumination – *negative* (cf. Ranula).

Differential diagnosis:

Ranula – transillumination positive.

(b) Inframylöhoid variety or cervical variety

1. *Midline cystic swelling just below the symphysis menti* (i.e., in the submental region). The swelling is sometimes so prominent that the patient seeks advice because of ‘double chin’ appearance.
2. Does not move with deglutition (cf. Thyroid swelling).
3. Does not move on protruding the tongue (cf. Thyroglossal cyst).
4. Fluctuation – *positive*.
5. Transillumination – *negative*.
6. Bimanually palpable with one finger in the mouth and one beneath the chin.

Differential diagnosis:

1. Ectopic thyroid
2. Suprahyoid thyroglossal cyst
3. Submental lymph node swelling.

II. Lateral variety

(a) Supramylöhoid or sublingual

- Cystic swelling in the floor of the mouth placed laterally.
- Transillumination – negative (cf. Ranula).

(b) Inframylöhoid or cervical

- Cystic swelling in the region of the submandibular salivary gland.
- Transillumination – negative.

Differential diagnosis:

1. Plunging ranula: Bidigitally palpable; transillumination is positive.
2. Submandibular salivary gland swelling – solid swelling.
3. Submandibular lymph node swelling – solid swelling.

Complications

1. Cosmetically looks ugly.
2. If too big, may cause difficulty in speech or eating, respiratory difficulty.
3. Infection.
4. Repeated trauma.

Treatment

Complete excision.

Sublingual dermoids are approached through the floor of the mouth.

Cervical dermoids are approached via a curved incision along the Langer's line over the submental or submandibular region. Sometimes the dissection is to be carried out from the neck into the oral cavity by retracting the posterior border of the mylohyoid muscle.

THYROGLOSSAL CYST

It is a cystic swelling arising from the remnant of the thyroglossal duct. It is regarded as tubulodermoid.

Surgical anatomy

The thyroglossal duct extends as a downgrowth of solid column of cells from the foramen caecum to the isthmus of the thyroid gland. Subsequently the solid column undergoes canalisation. This duct ultimately disappears when its function is over. The other name of the duct is *median thyroid diverticulum*.

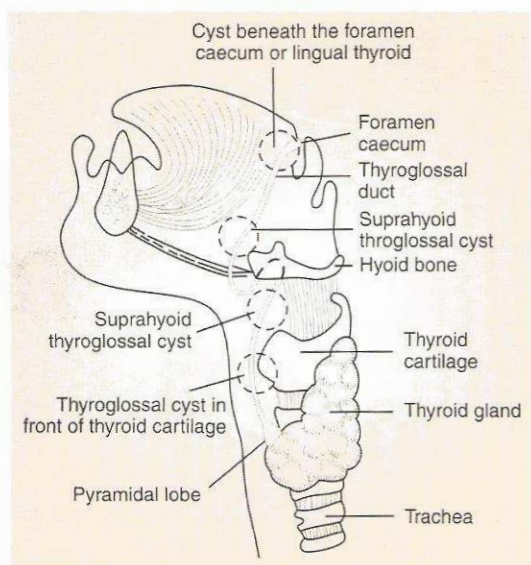


Fig. 3.7. The course of the thyroglossal duct and different sites of the thyroglossal cyst.

Path of the duct

It starts from the foramen caecum of the tongue and descends through the genioglossi muscles up to the hyoid bone (Fig. 3.6). At the region of the hyoid bone it descends:

- either in front of the hyoid bone, or
- through the hyoid bone, or
- it hooks below and behind the hyoid bone (Fig. 3.7).

Then it descends up to the level of the upper border of the thyroid cartilage.

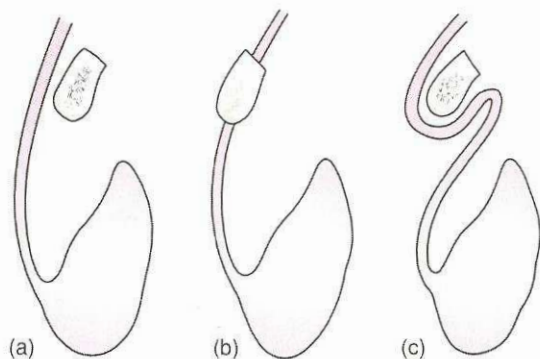


Fig. 3.8. Relation of the thyroglossal duct with the hyoid bone: (a) in front of the hyoid bone, (b) through the hyoid bone, and (c) hooks below and behind the hyoid bone.

Fate of the duct

- It completely atrophies except at the lower part where it forms the *isthmus of the thyroid gland*, and the *pyramidal lobe*, if present.
- Part from foramen caecum up to the hyoid bone disappears. The rest persists as *levator glandulae thyroidae*.
- From the portion of the diverticulum ectopic thyroid tissues may develop which in adult life are situated along the course of the diverticulum. When present in the region of the tongue, it is called *lingual thyroid*. A *lingual thyroid* looks like a flattened strawberry sitting on the base of the tongue. It may be the only thyroid tissue present in the body.
- A portion of the duct may remain unobliterated and gives rise to a cystic swelling due to accumulation of secretions—*thyroglossal cyst*.

Sites where thyroglossal cyst may occur

Anywhere along the course of the duct (Fig. 3.7), e.g.

- In the substance of the tongue beneath the foramen caecum (rarest).
- In the floor of the mouth.
- Suprahyoid region.
- Subhyoid region (commonest).
- In front of the thyroid cartilage.
- In front of the cricoid cartilage.

Pathology

Lining of the cyst wall: Cyst is lined by columnar, cuboidal or squamous epithelium and surrounded by a shell of lymphoid tissue (so, prone to infection). It may contain thyroid tissue.

Contents: Transparent, thick, jelly-like fluid; cholesterol crystal may be present. Occasionally clotted blood may be present.

Diagnostic criteria

- Age:** Commonly found in children but may occur at any age.
- Sex:** More common in women.
- Midline cystic swelling** (Figs. 3.9, 3.10A & B): Thyroglossal cyst usually presents as an asymptomatic swelling in the upper part of the neck in the midline. It is usually present around the hyoid bone. Cyst in front of thyroid cartilage

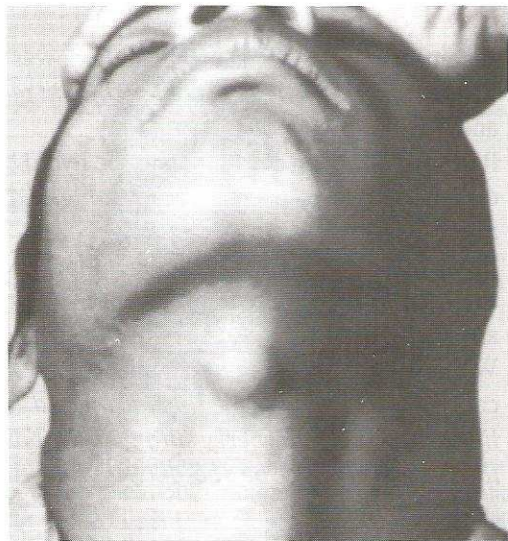


Fig. 3.9. Thyroglossal cyst. Midline cystic swelling in front of the hyoid bone.

is situated slightly lateral to the midline, usually to the left.

4. Usually elongated in shape having its long axis along the long axis of the neck (cf. Subhyoid bursal cyst). The size varies from 0.5 to 5 cm in diameter.
5. *Moves with deglutition* (because the swelling is attached to hyoid bone by fibrous tissue).
6. *Swelling also moves up on protrusion of the tongue* (because the swelling is attached to the tongue by the persistent obliterated thyroglossal duct).
7. The swelling can be moved sideways, but not up and down. Surface is smooth and the skin over the swelling is free.
8. *Cystic in feel* but fluctuation is difficult to elicit, because it contains thick glairy fluid under tension. Paget's test may help.
9. Transillumination is negative but rarely may be positive.
10. Usually painless and not tender unless infected.
11. Regional lymph nodes are usually not enlarged.

Differential diagnosis

1. Subhyoid variety is to be differentiated from the subhyoid bursal cyst. *Thyroglossal cyst moves up on protrusion of the tongue but not the subhyoid bursal cyst.*

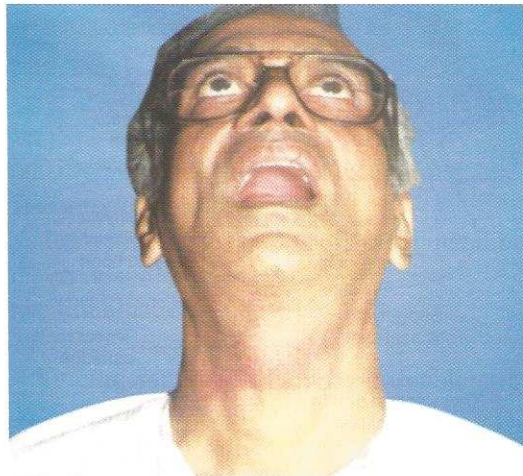


Fig. 3.10A. Thyroglossal cyst. Midline cystic swelling in front of upper border of thyroid cartilage.

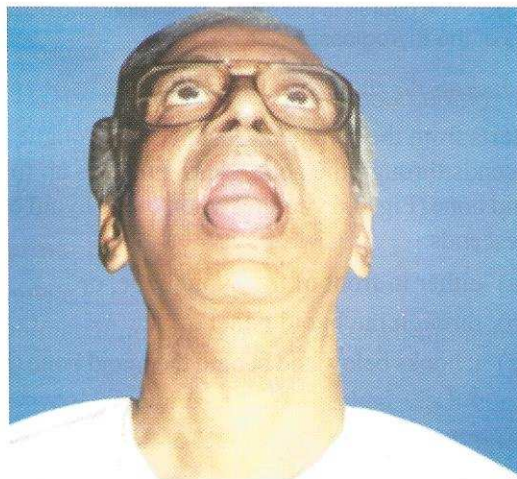


Fig. 3.10B. Same patient. On protrusion of the tongue the swelling moves up.

2. Suprahyoid variety is to be differentiated from the cervical dermoid cyst situated below the mylohyoid (inframyllohyoid) – same as above.
3. Thyroglossal cyst in front of thyroid or cricoid cartilage is to be differentiated from a cystic swelling in connection with the thyroid gland particularly related to the isthmus of thyroid. In both the conditions the swelling moves up with deglutition. But on protrusion of the tongue the thyroglossal cyst only moves up but not the cyst in connection with the thyroid gland.

What are the swellings that move with deglutition?

1. Thyroid swellings.
2. Cyst in relation to the isthmus of the thyroid gland.
3. Ectopic thyroid.
4. Thyroglossal cyst.
5. Subhyoid bursal cyst.
6. Enlarged pretracheal glands.
7. Laryngocele.

But only the thyroglossal cyst moves up also on protrusion of the tongue.

Complications

1. Recurrent infection (because the cyst is surrounded by a shell of lymphoid tissue).
2. Fistula formation.
3. Malignancy – Papillary carcinoma can occur rarely in the thyroglossal duct remnants.

THYROGLOSSAL FISTULA

Aetiology

1. *Congenital*: Rare, when the cellular median thyroid diverticulum communicates with the skin of the neck.
2. *Mostly acquired* because of:
 - bursting of the infected thyroglossal cyst;
 - inadvertent incision over the infected cyst mistaking it for an abscess;
 - incomplete removal of the thyroglossal cyst.

Though the fistula is mostly acquired, the persistence of the tract is always a congenital anomaly.

Pathology

Lining of the tract: Usually by squamous cell epithelium.

Nature of the discharge: Serous or mucoid; if infected, purulent.

Diagnostic criteria

1. Usually found in children or young adults. Rarely may be present at birth. (So, ask about duration — congenital or acquired.)
2. *Small fistula's opening in the midline of the neck* usually below the hyoid bone hence also known as *median fistula of the neck* (cf. Branchial fistula — which is known as lateral fistula of the neck)..

3. The fistula may be infected with purulent discharge. In the quiescent stage there may be mucoid discharge.
4. *The fistula's opening moves up on protrusion of the tongue* (because of the persistence of the tract which is attached to the base of the tongue).
5. *Characteristic feature is the hooding of the skin above the opening*, i.e., a crescentic fold of skin with the concavity downwards, overlying the fistula's opening. This hooding of the skin becomes more prominent on protrusion of the tongue.
6. The fistula may close intermittently with recurrence of pain and inflammation.
7. Enquire about any previous swelling which might have burst or been incised.

Treatment of thyroglossal cyst and fistula

Complete excision of the cyst, or of the fistula tract with the removal of every vestige of the thyroglossal tract as close as possible to the base of the tongue. Otherwise there may be recurrence of thyroglossal fistula.

Sistrunk's operation

Where, in addition to the above measures, a portion (particularly the central) of the hyoid bone is excised. For, the body of the hyoid bone may obstruct the dissection particularly when the tract is hooking below and behind the hyoid bone or passing through its substance during its descent.

Pre-operative precautions and measures

1. The suspected swelling may be due to ectopic thyroid tissue. So, before excision, if it is doubtful, one should confirm that it is not the only thyroid tissue present in the body, otherwise its excision will be followed by hypothyroidism. *So, in case of any doubt, it is to be confirmed by radioactive iodine uptake test.*
2. For better visualisation of the tract, methylene blue may be injected into the cyst. When the fistula is present, the dye is pushed through the fistula's opening on the night before operation, and the opening of the fistula is closed with a purse-string suture. This procedure helps in better visualisation of the tract during operation.
3. If there are any features of infection, they should be controlled with antibiotics prior to surgery.

THYROGLOSSAL FISTULA

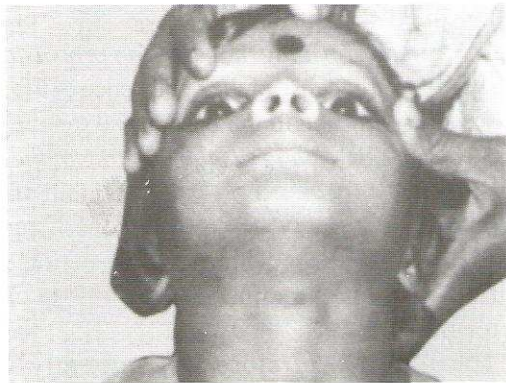


Fig. 3.11A. Thyroglossal fistula—midline fistula. So, known as the median fistula of the neck.



Fig. 3.11B. Same patient. Fistula moves up on protrusion of the tongue.



Fig. 3.12. Recurrent thyroglossal fistula. Most probably due to inadequate excision. Note the previous scar mark below the fistula. Sistrunk's operation is always advisable.

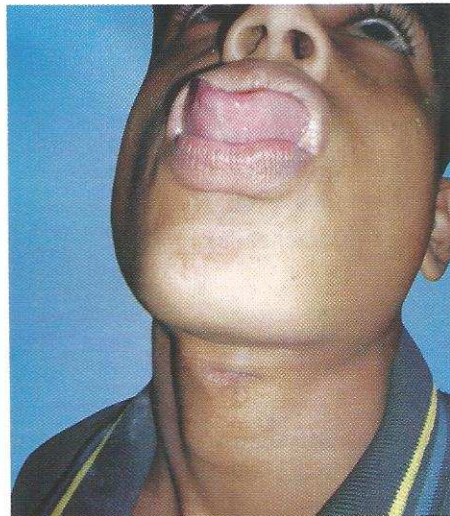


Fig. 3.13. Recurrent thyroglossal fistula—moved up on protrusion of the tongue. Note the scar mark from previous operation. Sistrunk's operation is always advisable. (Courtesy: Prof. Sudeb Saha, MS, FAMS)

SUBHYOID BURSAL CYST

It is the accumulation of inflammatory fluid giving rise to a cystic swelling of the subhyoid bursa.

The subhyoid bursa is situated over the thyrohyoid membrane below the hyoid bone slightly posterior to its lower border.

Pathology

Lining of the cyst — epithelium.

Contents — turbid or clear fluid.

Diagnostic criteria

1. History of painful onset with features of mild inflammation.
2. Midline swelling but transversely elongated — discoid-shaped cystic swelling (cf. Thyroglossal cyst).
3. Situated below the hyoid bone.
4. Moves with deglutition (as attached to hyoid bone).
5. But no movement on protrusion of the tongue (cf. Thyroglossal cyst).
6. Cystic in feel — fluctuation is positive.
7. Transillumination — usually negative; may be positive if fluid is clear.

Treatment

Complete excision by skin crease incision.

BRANCHIAL CYST AND FISTULA

Before considering the cysts and fistula, a short account of branchial arches and their derivatives is presented here.

DEVELOPMENT OF BRANCHIAL ARCHES

In the developing embryo surrounding the primitive pharynx (or cephalic part of the foregut) six pairs of mesodermal condensation appear in the form of branchial or visceral arches, derived from the lateral sheet mesoderm (Fig. 3.14).

In between the arches, there are depressions or spaces both inside and outside the pharynx. Internally these spaces are lined by entoderm and are called branchial or pharyngeal pouches. Externally these spaces are lined by ectoderm and are called branchial or pharyngeal clefts. So, between the two is the cleft membrane, which is lined internally with columnar and externally with squamous epithelium. The cleft membrane never disappears in human body (in contrast to fishes), because a little mesodermal element always persists between the entoderm and ectoderm.

Each branchial arch has a central plate of cartilage (which later gives rise to bone), a muscle mass, a nerve (to supply the derivatives of the arch) and an artery.

Mesodermal derivatives of the branchial arches**First arch or mandibular arch****A. From the cartilage:**

1. Malleus
2. Incus
3. Lingula of the mandible
4. Mental ossicles, if present.

B. From the sheath around the cartilage:

1. Anterior ligament of malleus
2. Sphenomandibular ligament
3. Body and ramus of the mandible.

C. From the muscle mass or myotome:

All the muscles supplied by mandibular nerve:

1. Muscles of mastication — masseter, temporal, lateral and medial pterygoids
2. Tensor palati
3. Tensor tympani
4. Mylohyoid
5. Anterior belly of digastric.

D. Nerve of the first arch:

- Mandibular nerve (a branch of trigeminal nerve or 5th cranial nerve).

E. Artery of the first arch:

- Maxillary artery (remnant of the 1st aortic arch).

Second arch or hyoid arch**A. From the cartilage (Reichert's cartilage):**

1. Stapes
2. Styloid process of the temporal bone
3. Lesser cornu and upper half of the body of the hyoid bone.

B. From the sheath around the cartilage:

- Stylohyoid ligament.

C. From the muscle mass or myotome:

All the muscles supplied by the facial nerve:

1. Muscles of facial expression including occipitofrontalis and buccinator
2. Platysma
3. Posterior belly of digastric
4. Stylohyoid
5. Stapedius.

D. Nerve of the 2nd arch:

- Facial nerve (7th cranial nerve).

E. Artery of the 2nd arch:

- Stapedial artery (remnant of the dorsal part of 2nd aortic arch).

Third arch**A. From the cartilage:**

- Greater cornu and lower half of the body of the hyoid bone.

B. From the muscle mass or myotome:

- A muscle supplied by the glossopharyngeal nerve — stylopharyngeus muscle.

C. Nerve of the 3rd arch:

- Glossopharyngeal nerve (9th cranial nerve).

D. Artery of the 3rd arch:

- Internal carotid artery (remnant of the dorsal part of 3rd aortic arch); common carotid artery (remnant of the ventral part of the 3rd aortic arch).

Fourth arch

A. From the cartilage:

1. Thyroid cartilage
2. Epiglottis.

B. From the muscle mass or myotome:

All the muscles supplied by the external laryngeal nerve which is a branch of superior laryngeal nerve.

1. Cricothyroid
2. Inferior constrictor of pharynx.

C. Nerve of the 4th arch:

- Superior laryngeal nerve – a branch of vagus nerve.

D. Artery of the 4th arch:

1. Right side – first part of the subclavian artery.
2. Left side – main part of the arch of the aorta (both are remnants of the 4th aortic arch).

Fifth arch

- Disappears.

Sixth arch

A. From the cartilage:

1. Cricoid cartilage, arytenoid cartilage (probably corniculate and cuneiform cartilages).
2. Rings of trachea and bronchi.

B. From the muscle mass or myotome:

All the muscles supplied by the cranial accessory nerve carried via the recurrent laryngeal nerve.

1. All the intrinsic muscles of the larynx except the cricothyroid.
2. All the muscles of the pharynx except the stylopharyngeus.
3. All the muscles of the palate except the tensor palati.

C. Nerve of the 6th arch:

- Recurrent laryngeal nerve – a branch of the vagus nerve (10th cranial nerve) but carrying the fibres of cranial accessory nerve.

D. Artery of the 6th arch:

1. Ventrally – main part of the pulmonary artery.
2. Dorsally – on the right side the artery

disappears; on the left side the artery persists as the ductus arteriosus (both are the remnants of the 6th aortic arch).

BRANCHIAL CYST

It is a cystic swelling arising in connection with the persistent cervical sinus which is formed due to the fusion of overgrowing 2nd branchial arch with the 6th branchial arch.

Pathogenesis

Conventional theory (Fig. 3.14)

The 2nd branchial arch migrates towards the surface and overgrows the 3rd and 4th arches (Fig. 3.14A) and ultimately fuses with the 6th arch forming a cavity called cervical sinus (5th arch disappears completely). Normally this sinus disappears. But if it persists, accumulation of fluid occurs inside the

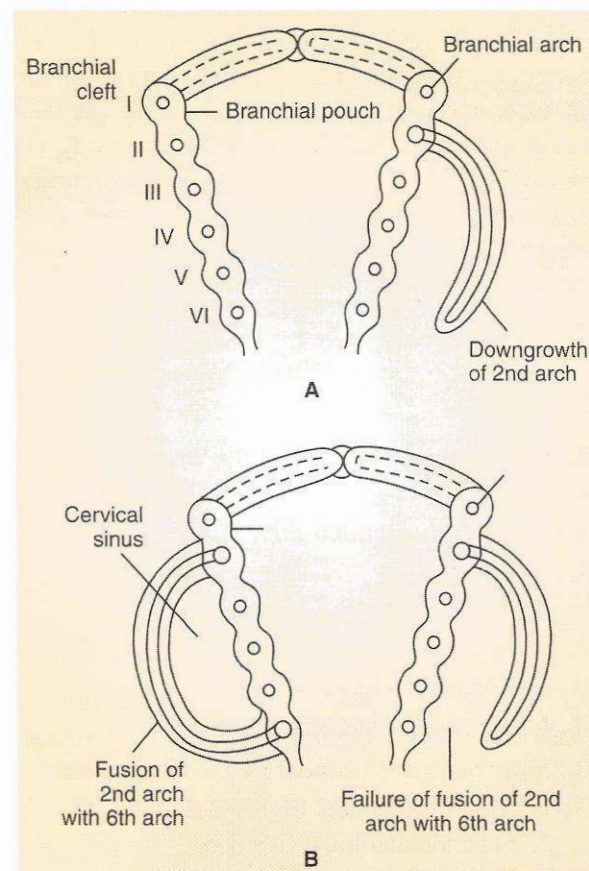


Fig. 3.14. The method of formation of branchial cyst and branchial fistula.

sinus and gives rise to a cystic swelling called branchial cyst. The secretion usually comes from the appendages of the ectodermal lining of the enclosed space, sweat and sebaceous glands.

The branchial cyst usually lies superficial to the structures derived from the 2nd and 3rd branchial arches, i.e., the lesser cornu of hyoid bone, posterior belly of digastric muscles, facial nerve, external carotid artery.

Sometimes the 2nd arch fails to fuse with the 6th arch and thus gives rise to branchial sinus or fistula (Fig. 3.14B).

Other theories of origin of branchial cyst

1. One theory suggests that the cervical sinus is closed from the bottom to the surface and during this process ectodermal epibranchial placodes grow inwards. The placodal vesicles usually disappear after organising the development of the ganglion connected with some cranial nerves. If the placodal vesicles persist they are expressed as branchial cyst.
2. It has been suggested that the branchial cyst may develop from the inclusion of the parotid epithelium in the upper deep cervical lymph nodes (*Illingworth*) (cf. Warthin's tumour).

Pathology

Lining of the cyst wall: Squamous epithelium (rarely columnar) surrounded by a large amount of lymphoid tissue, so prone to infection.

Contents: Cheesy material. It may resemble the appearance of cold abscess. But its characteristic feature is the presence of cholesterol crystals in large numbers.

Diagnostic criteria

1. **Age:** Although congenital, it first appears around 20 and 25 years, but its advent may be postponed till the 50th year. (Why? Because the fluid which it contains accumulates very slowly.)
2. **Sex:** Equally common in both sexes.
3. The patient presents with a slow growing painless lump in the upper lateral part of the neck.

Examination:

4. **Site:** The lump lies below the angle of the jaw, beneath the middle of the sternomastoid muscle

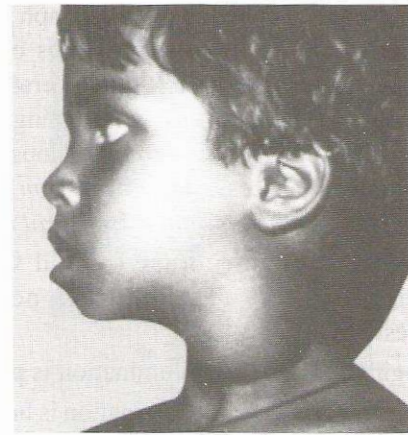


Fig. 3.15. Branchial cyst. A cystic swelling at left carotid Δ below the angle of the mandible. Posterior margin of the swelling overlapped left sternomastoid. Transillumination negative. Here the age of presentation is 10 years.

and bulges forward around the anterior border of the muscle into the carotid triangle. Very rarely the cyst can bulge backwards behind the muscle.

5. **Shape and size:** Usually ovoid and varies from 5 to 10 cm in width.
6. **Surface:** Smooth and the edge is distinct. Overlying skin is free and looks normal unless infected.
7. The lump is a tense, cystic swelling. *Fluctuation is positive.* The fluctuation is not always easy to elicit, specially when the cyst is deep to the upper part of the sternomastoid muscle. The lump is usually non-tender unless infected.
8. The lump is not very mobile as it is closely tethered to the surrounding structures.
9. **Transillumination:** Negative because of its thick contents.
10. **Lymph nodes:** The local deep cervical lymph nodes should not be enlarged. If they are, one should reconsider the diagnosis in favour of cold abscess rather than a branchial cyst.
11. **On aspiration:** The aspirated materials resemble the appearance of cold abscess. But the aspirate may demonstrate fat globules, and cholesterol crystals secreted by the sebaceous glands in the epithelial lining.

Differential diagnosis

1. Cold abscess in the neck: A caseating tuberculous lymph node may simulate a branchial cyst. But

the presence of enlarged matted lymph nodes in the neck along with other features of tuberculosis, such as evening rise of temperature, loss of weight, anorexia, excessive sweating etc., are helpful in the diagnosis of tuberculous lymphadenitis with abscess. *Aspiration from the cold abscess does not show any cholesterol crystals.*

2. Cervical dermoid (inframylahyoid variety): Aspiration from dermoid does not show cholesterol crystals.
3. Plunging ranula: Transillumination is positive.
4. Cystic hygroma: Transillumination is brilliantly positive and commonly situated in the posterior triangle.
5. Carotid body tumour – Solid swelling.
6. Solitary enlarged cervical lymph node – Solid swelling.
7. Submandibular salivary gland swelling – Solid swelling.

Complications

1. Recurrent infection (because of the presence of lymphoid tissue in the wall).
2. Followed by bursting – resulting in fistula formation.

When there is infection of the branchial cyst it may look like a hot abscess and when it is inadvertently incised branchial fistula is formed.

Treatment

Excision of the cyst through incision along the Langer's line.

Precautions during operation

1. The cyst wall is thin and delicate – so not to be held by any tissue forceps.
2. Avoid injury to the following structures:
 - Hypoglossal and spinal accessory nerves.
 - Carotid arteries.
 - Wall of the pharynx.

BRANCHIAL FISTULA

This may be either congenital or acquired.

1. *Congenital*: Commonest. It is due to either:
 - failure of fusion of the 2nd branchial arch with the 6th branchial arch (Fig. 3.12); or

- disappearance of the membrane (cleft membrane) connecting these two arches.

(Some others suggest thymopharyngeal duct as the origin of branchial fistula.)

2. *Acquired*: It is due to either:

- bursting of the infected branchial cyst; or
- inadvertent incision over the infected branchial cyst when mistaken for an abscess.

Types of congenital variety of branchial fistula

1. *Incomplete*: Commonest. It does not communicate with the cavity of the pharynx and remains blind at the inner end against the lateral pharyngeal wall. So, this type should better be termed branchial sinus.
2. *Complete variety* (Fig. 3.14): In this variety both external opening and internal opening are present, and hence the branchial fistula. The internal opening may open inside the pharynx either at the intratonsillar cleft or in the pyriform fossa.

Considering the above discussion, theoretically, three varieties of branchial fistulae may be recognised:

- (i) A complete fistula with an external opening on the skin, and an internal opening in the pharynx (Fig. 3.16). This is usually the result of inadvertent trauma during exploration of the tract by a probe which ruptures the tridermal-cleft membrane, separating the tract from the pharynx (McGregor).
- (ii) Only an internal opening in the pharynx (only theoretically possible).
- (iii) Only an external opening on the skin (commonest).

Path of the fistula tract

The superficial and deep relations of a branchial fistula are the same as in the case of a branchial cyst.

Traced from below, the tract pierces the deep fascia at the upper border of thyroid cartilage – then passes between the fork of the common carotid artery lying superficial to internal carotid artery (3rd arch derivative) but deep to external carotid artery and its branches (Fig. 3.16). When traced further towards the pharynx, the tract lies superficial to stylo-

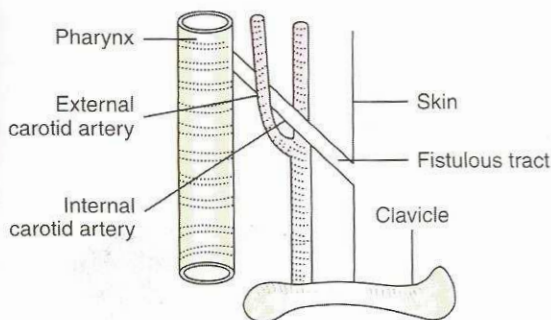


Fig. 3.16. The course of a branchial fistula—a complete variety.

pharyngeus muscle, glossopharyngeal nerve (3rd arch derivative) but deep to the hypoglossal nerve and the stylomandibular ligament.

Pathology

Lining wall of the sinus: Squamous epithelium or pseudostratified ciliated columnar epithelium deep to which is a layer of lymphoid tissue.

Nature of discharge: Mucoïd or mucopurulent with or without excoriation of the surrounding skin.

Diagnostic criteria

1. May be found at any age but usually seen in growing adults.
2. Equally common in male and female.
3. May be unilateral or bilateral.
4. *Site of external opening:* In congenital variety a small dimple or an external opening is situated at the typical site, i.e., at the anterior border of sternomastoid at the junction of the lower 1/3rd & the upper 2/3rd (cf. Branchial cyst) (Fig. 3.17). But in the acquired variety, the dimple will be at a higher level, i.e., at the level of branchial cyst. Branchial fistula is also called *lateral fistula of the neck* (cf. Thyroglossal fistula—which is called median fistula of the neck).
5. Swallowing may accentuate the dimple on the skin.
6. Mucoïd or mucopurulent discharge.
7. Excoriation of surrounding skin may be present.
8. Branchial cartilage may be present on palpation.

Treatment

Complete excision of the tract after thorough dissection starting from the cutaneous opening up to the wall of the pharynx.

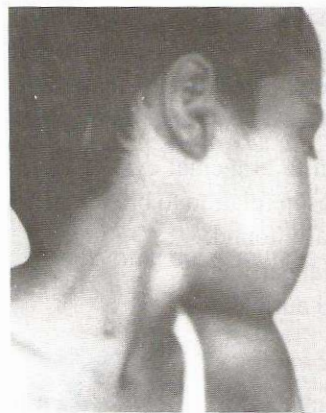


Fig. 3.17. Branchial fistula. Note the external opening at the junction of lower 1/3rd and upper 2/3rd of the sternomastoid muscle (cf. Branchial cyst).

Pre-operative measures

Sinogram: Injection of radio-opaque dye into the fistulous tract followed by straight X-ray in order to assess the upper limit and extent of the fistula. This may be of some help during excision of the tract.

Steps of operation

1. Incision: An elliptical incision along the Langer's line encircling cutaneous opening of the fistula.
2. After cutting superficial fascia, platysma, the tract is dissected from below up as high as possible.

Gentle traction on the tract, by pulling the excised cutaneous opening, facilitates identification and proximal dissection of the tract.

3. Sometimes a second incision is needed above the upper border of thyroid cartilage. This second incision is made parallel to the first incision and the dissected tract is taken out through the second incision. This is known as '*step-ladder pattern*' of dissecting a branchial fistula.

It is generally possible to trace the fistula up to the lateral wall of the pharynx, where it is ligated and excised.

4. After removal of the tract, the skin is closed with or without a drain.

Precautions during operation

1. As the tract passes through the bifurcation of the common carotid artery, the vessels should be protected.

2. Care should be taken not to injure the hypoglossal nerve which lies superficial to the tract.
3. Complete removal is necessary, otherwise persistence of the tract will cause recurrence.

In addition to branchial cyst or fistula, two other abnormalities in relation to branchial apparatus may be occasionally found:

- (a) Branchial cartilage
- (b) Cervical auricle.

Branchial cartilage

It is an elongated piece of cartilage embedded in the skin, situated deep to the cutaneous dimple in relation to the external opening of the branchial fistula.

Cervical auricle

A hood of skin looking like an ear, containing yellow or elastic fibrocartilage, is found in relation to the external opening of the branchial fistula.

What is branchiogenic carcinoma?

It is a rare neoplasm arising from the remnants of the branchial clefts. The condition is very rare. Existence of a proved case is extremely doubtful.

CYSTIC HYGROMA

A cystic hygroma is a collection of lymphatic sacs containing clear colourless lymph.

It is a congenital malformation affecting the lymphatic channels and arises from the congenital lymph sacs which are the precursors of adult lymphatic channels. It is a variety of lymphangioma, and also regarded as hamartoma.

Pathogenesis

At the 6th week of intrauterine life 3 pairs of lymph sacs appear in the embryo from which the adult lymphatic channels are derived.

- One pair in the neck — jugular lymph sacs.
- One pair in the retroperitoneum.
- One pair near the inguinal region below the bifurcation of the common iliac vein — posterior lymph sacs.

By and large, cystic hygromas develop from these primitive sacs. *In the neck, cystic hygroma develops from the remnants of the jugular lymph sacs.*

The jugular lymph sacs in the neck appear one on

either side, at the junction of the internal jugular vein and subclavian vein (Fig. 3.18). In the lower animals these lymph sacs are called primitive lymph heart.

During development, a portion of this lymph sac along with its lymphatic channels may be sequestered into the surrounding mesoderm (Fig. 3.18). This sequestered portion of the lymph sac ultimately loses its communication with the primitive lymph sac.

Later, this isolated portion of the lymph sac lined by columnar epithelium secretes lymph and gets distended to give rise to a lymphatic swelling known as cystic hygroma.

Pathology

1. It appears very early and may be found at birth or within the first few years of life. Of all the congenital swellings it is the earliest to develop, even before sternomastoid tumour.
2. It consists of multiple cysts—so, multilocular and multilobular.
3. The cysts are filled up with clear lymph.

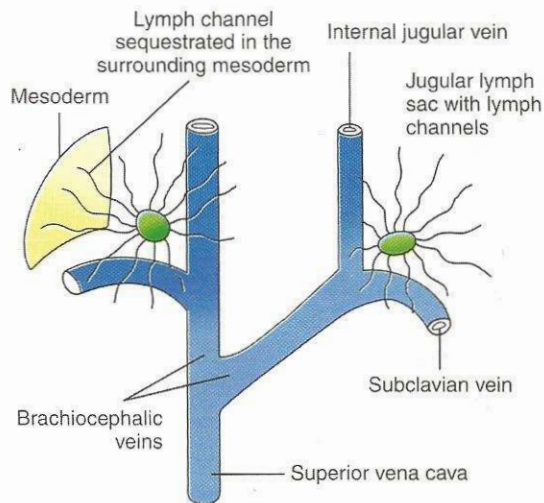


Fig. 3.18. The method of formation of cystic hygroma.

4. The larger cysts are situated at the periphery and the smaller cysts at the centre.
5. Many of the cysts may intercommunicate with one another.
6. Though cystic hygromas infiltrate into the surrounding tissues, *they never turn malignant.*
7. *Lining of the cyst:* Each cyst is lined by a single layer of columnar epithelium and covered externally with a shell of lymphoid tissue.

8. *Contents*: Clear and watery or straw-coloured fluid consisting of cholesterol crystals, lymphocytes. The fluid does not coagulate.

Usual sites of cystic hygroma

1. Root of the neck in the posterior triangle – commonest site. It may extend upwards towards the ear or downwards towards the axilla and/or mediastinum.
2. Axilla.
3. Groin or inguinal region.
4. Mediastinum.
5. Tongue and buccal mucosa of the cheek. Usually lymphangiogenetic macroglossia occurs in connection with the cystic hygroma.

So, these sites are also to be examined during examination of a cystic hygroma.

Diagnostic criteria

1. *Age*: Infants or young children.
2. *Site*: Commonly at the root of the neck in the posterior triangle, deep to the sternomastoid (Figs. 3.19, 3.20).
3. The swelling increases in size and becomes prominent when the child cries or strains (due to rise of intrathoracic pressure which is transmitted through the root of the neck). This feature may be informed by the parents or can be observed (Fig. 3.21A & 3.21B).
4. *Soft cystic swelling—fluctuation positive*.
5. Surface is *lobulated*. The overlying skin is free and normal unless infected.
6. Margins are not well-defined on all sides, as it burrows into the tissue space.
7. Sometimes partially compressible (because of intercommunication).
8. *Transillumination—brilliantly positive*, a distinctive physical sign. During transillumination multiple septae are noticed—*multilocular cyst*.
9. Regional lymph nodes are not enlarged.
10. Other sites usual for the cystic hygroma are also to be examined (e.g., axillae, groin).

Atypical presentation of cystic hygroma

1. During birth a big cystic hygroma may cause obstructed labour.

2. Due to recurrent infection it may present as a pyogenic abscess.
3. Cystic hygroma in the mediastinum may present as a growth in the mediastinum with mediastinal syndrome characterised by dyspnoea, dysphagia, etc. Clinically it is very difficult to differentiate mediastinal cystic hygroma from a mediastinal tumour. Many a time the dispute is settled at the operation table.

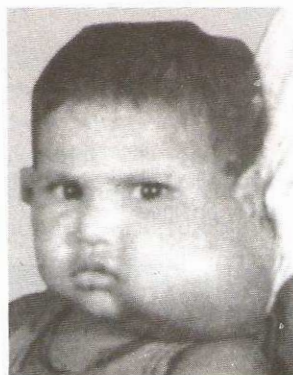


Fig. 3.19. Cystic hygroma of the left neck extending to the face. Transillumination was highly positive.

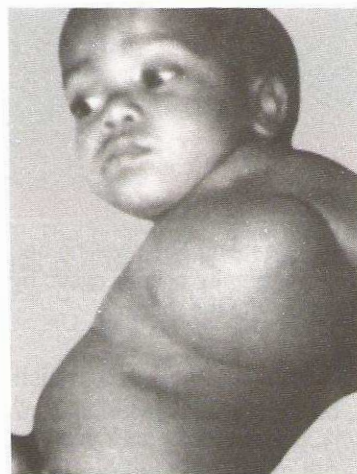


Fig. 3.20. Cystic hygroma involving neck and axilla. Axillary swelling was very big in this case. So, always examine the axillae, groins in addition for a second swelling in a patient with cystic hygroma.

Differential diagnosis

1. Branchial cyst.
2. Cold abscess in the neck.
3. Sometimes a collar-stud abscess.



Fig. 3.21A. Cystic swelling at the root of the neck. Transillumination is highly positive.

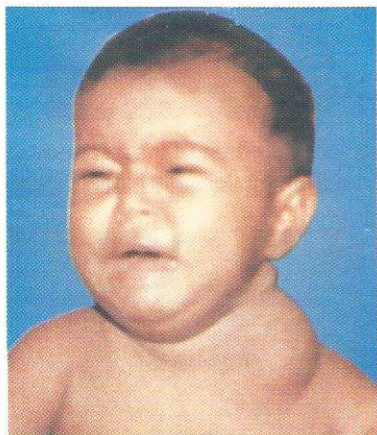


Fig. 3.21B. Same patient. The swelling becomes more prominent when the baby cries – cystic hygroma with possible extension into the thorax.

In all the above mentioned cases transillumination is negative, but *in cystic hygroma transillumination is brilliantly positive.*

4. *Solitary lymph cyst:* This is a *single cyst* containing lymph and develops in the same process as cystic hygroma. Though congenital, it usually *appears in adult life* and is commonly found in the supraclavicular region (Fig. 3.22).

Complications

1. Recurrent infection – because the cyst is surrounded by a shell of lymphoid tissue.
2. Sudden increase in the size of the cyst may cause respiratory distress – when the treatment is



Fig. 3.22. A cystic swelling in the supraclavicular region. Brilliantly translucent cyst without any septae within (i.e., not multiloculated)—Solitary lymphatic cyst.

aspiration of the cyst with or without tracheostomy.

3. Never turns into a malignancy.

Can spontaneous recovery take place?

Yes, and when it occurs it will take about 2 years to recover.

Treatment

1. If there is no urgent indication, the operation can be delayed, and the child is observed for 2 years, because there is a chance of spontaneous recovery.
2. Before commencing any treatment, X-ray of the chest is to be done to see any evidence of mediastinal cystic hygroma. X-ray may reveal shadow at the hilar region or in any part of the mediastinum.

3. Conservative:

- (a) Aspiration alone, particularly when the cyst is big enough to cause pressure symptoms.

Fallacy:

- There are multiple cysts with intercommunication between them. Therefore, single aspiration cannot bring about permanent cure.
 - Repeated aspiration leads to infection.
- (b) Aspiration followed by injection of hot water or hypertonic saline.

Advantage: Injection of sclerosing agents causes fibrosis with the following advantages:

- the size diminishes;
- the cyst becomes more localised;
- dissection is easier because the cyst wall becomes thick.

4. **Operative:** Complete excision of the cyst is the treatment of choice.

Precautions during excision:

- (a) Cyst wall should never be held with tissue forceps because the wall is very thin, delicate and it will give way — so chances of incomplete removal.
- (b) There are finger-like projections from the cyst wall invading the surrounding structures. So, every care should be taken for complete excision.

Dangers of incomplete removal:

- Fluid and electrolyte loss leading to dehydration may rarely develop, which is very difficult to correct in a child.
- Wound infection.
- Recurrence.

Sometimes, thoracotomy may have to be done for mediastinal extension of cystic hygroma. Occasionally, tracheostomy may have to be done if there is prolonged tracheal compression.

5. **Radiotherapy:** Cystic hygroma is relatively radio-resistant. But it may be used in case of recurrence, or when the surgical treatment is not feasible.

Name the cysts containing cholesterol crystals

1. Branchial cyst
2. Cystic hygroma
3. Thyroglossal cyst
4. Dental cyst
5. Dentigerous cyst
6. Old hydrocele — occasionally.

PHARYNGEAL POUCH

It is a protrusion or herniation of the mucosa of the pharyngeal wall through the *Killian's dehiscence*. Truly, it is a pressure diverticulum or *Pulson diverticulum of the pharynx*.

Surgical anatomy

Killian's dehiscence (Fig. 3.23)

The inferior constrictor muscle has got two parts:

- (i) upper oblique fibres (thyropharyngeus), and
- (ii) lower horizontal fibres (cricopharyngeus).

In between these fibres on the posterior aspect of pharyngeal wall there is a potential area of weakness which is known as '*Killian's dehiscence*'. At this site often there is a depression called pharyngeal dimple.

Both these fibres of inferior constrictor are different in their arrangements of fibres, nerve supply and functions.

Arrangement of fibres

Thyropharyngeus: The fibres are oblique in direction and are inserted into the median raphe. This median raphe does not extend below the level of vocal cord.

Cricopharyngeus: The fibres are horizontal in direction, around the upper end of the oesophagus and are continuous with the similar fibres of the opposite muscle. No median raphe exists here.

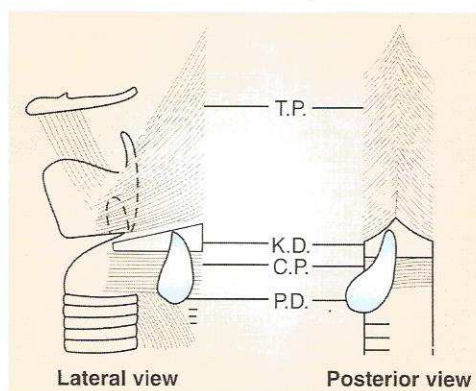


Fig. 3.23. The pharyngeal diverticulum. T.P., thyropharyngeus part of inferior constrictor; K.D., Killian's dehiscence; C.P., cricopharyngeus part of the inferior constrictor; P.D., pharyngeal diverticulum.

Nerve supply

- **Thyropharyngeus:** Supplied by the pharyngeal plexus carrying the fibres from the cranial part of the accessory nerve.
- **Cricopharyngeus:** Supplied by the recurrent laryngeal and external laryngeal nerves.

Functions

- **Thyropharyngeus** is propulsive in function, i.e., it propels the food downward during deglutition.
- **Cricopharyngeus** is sphincteric in action and is normally kept closed except during the act of deglutition.

- *When thyropharyngeus contracts, cricopharyngeus relaxes and vice versa.*

Clinical basis of development of pharyngeal pouch

Due to neuromuscular imbalance when both the parts contract simultaneously, intrapharyngeal pressure rises and the mucous membrane bulges through the Killian's dehiscence forming a pharyngeal diverticulum. This is called Pulson diverticulum which is the beginning of pharyngeal pouch.

Course of the diverticulum

Initially it starts in the midline posteriorly. But posteriorly lies the rigid vertebral column. So, it deviates on either side, mostly to the left.

Diagnostic criteria

1. Common in middle and old age. More common in men.
 2. Symptoms vary according to the pathological stages.
 - *Stage I or stage of initial bulging:* There is formation of a small diverticulum and it is usually symptomless. There may be symptoms of foreign body sensation in the throat.
 - *Stage II or stage of well-formed diverticulum:* The diverticulum becomes larger and globular in shape; the mouth of the diverticulum is vertical giving rise to following symptoms:
 - Sense of suffocation, cough, mild dysphagia.
 - Regurgitation of food, during the next meal or when the patient turns from side to side.
 This may rarely cause aspiration pneumonia.
 - *Stage III or stage of big diverticulum:* The diverticulum becomes larger, the mouth being horizontal. The fundus of the sac becomes dependent and may cause extraneous pressure over the oesophagus.
- Patient complains of:*
- Gurgling sounds in the neck especially when the patient swallows (the diverticulum being filled up with food materials, liquid and air).
 - Dysphagia.
 - Visible swelling in the neck.
 - Aspiration of the contents of the pouch leading to features of aspiration pneumonia, lung abscess, mediastinitis.
 - Features of cachexia due to starvation.

- Some patients can reduce the pouch after meal.

3. *Site:* When presented with swelling the pharyngeal pouch appears *behind the sternomastoid muscle below the level of thyroid cartilage*. The pouch lies deep to the deep fascia, behind the sternomastoid muscle, and is fixed deeply.
4. Shape and size – variable.
5. Surface smooth and the edge is not palpable.
6. *Soft swelling and indentable. It can be compressed and sometimes emptied.*
7. Fluctuation may be positive but transillumination is negative.

Investigations

1. X-ray: A barium swallow study using a thin barium reveals compression of the posterolateral oesophageal wall and filling of the pouch. The contents of the pouch will spill into the oesophagus on compression of the swelling in the neck.
2. Oesophagoscopy — dangerous.

Treatment

- *Stage I:* Wait and watch.
- *Stage II and Stage III:* Excision of the diverticulum either after a feeding gastrostomy or without it. Excision of the pouch is followed by cricopharyngeal myotomy like Heller's operation. The hypertrophied circular muscle is divided vertically till the mucosa is reached.

LARYNGOCELE

It is a diverticulum containing air, formed due to herniation of laryngeal mucosa through the thyrohyoid membrane. It is mostly unilateral. It simulates cervical air pouches found in many mammals (e.g., monkeys) who inflate them voluntarily.

Diagnostic criteria

1. *Mostly acquired.* Commonly found amongst the professional trumpet players. May be found in glass blowers and in persons suffering from the chronic cough.
2. When the sac enlarges it gives rise to a visible lateral swelling in the neck over the thyroid cartilage.

3. The swelling becomes visible or prominent when the patient is asked to blow or breathe out against closed mouth and nose (Valsalva manoeuvre).
4. Boggy in feel.
5. May be resonant on percussion.
6. History of appearance and disappearance of the swelling.
7. This swelling moves up with the larynx on swallowing. This is quite diagnostic.
8. History of hoarseness of voice may be present.

Complications

1. Sudden death due to asphyxia from rapid distension of laryngocele — a rare phenomenon.
2. Infection leading to laryngopyocele.

Treatment

Complete excision of the sac after thorough dissection followed by invagination of the stump.

CAROTID BODY TUMOUR

It is a tumour arising from the chemoreceptor cells of the carotid body situated at the bifurcation of common carotid artery. The other name is *chemodectoma* or *potato tumour*.

Carotid body

1. The carotid body is a flattened brownish nodule, a few millimetres in diameter.
2. It is situated within the adventitious coat on the posterior aspect of the common carotid artery at or just below the bifurcation.
3. It consists of parenchyma cells embedded in a fibrous stroma richly supplied with vessels and nerves.

The parenchyma cells are of two types:

- (i) Chief cells containing granules of catecholamine type.
 - (ii) Sustentacular cells having long processes embracing adjacent capillaries.
4. It belongs to the chemoreceptor system.

Sites where other chemoreceptor apparatuses present in the body (Fig. 3.24)

1. *Aortic bodies*: These lie near the points of origin of the left coronary artery, the innominate artery,

bifurcation of the pulmonary trunk in relation to ligamentum arteriosum, arch of aorta.

2. *Glomus jugulare*: It lies in the adventitia of the bulb of the jugular vein.
3. *Glomus intravagale*: It is associated with the ganglion nodosum of the vagus nerve.
4. *Paraganglion tympanicum*: It lies along the tympanic ramus of the glossopharyngeal nerve.
5. In addition, chemoreceptor apparatus may be found:
 - in relation to inferior alveolar artery in the orbit;
 - in relation to femoral artery in the femoral canal;
 - in the small bowel mesentery;
 - in the retroperitoneum.

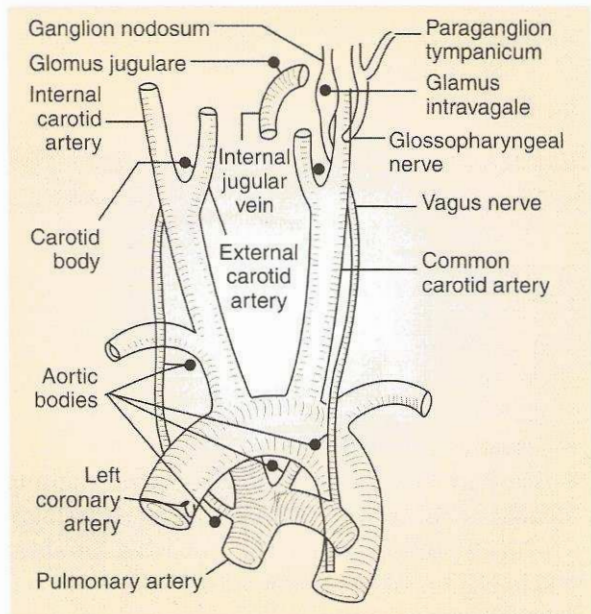


Fig. 3.24. The different sites of chemoreceptor.

Functions of chemoreceptors

The functions of the chemoreceptor cells are not clear. They are sensitive to the change of pH and carbon dioxide and oxygen tensions in the circulating blood. Lack of oxygen produces hyperplasia of these cells.

Pathology of carotid body tumour

1. It is regarded as non-chromaffin paraganglioma and is akin to glomus tumours of the skin and neuroblastoma of the sympathetic nervous system.

2. Previously it was thought to be benign. But cases have been reported with metastases. Regional metastases occur in about 20 percent cases but distant metastasis is negligible.
3. The tumour is hard and white-yellow, or spongy and vascular.
4. The tumour is well encapsulated with a yellow or orange cut surface and with dense fibrous septae.
5. Histology reveals solid masses resembling the chief cells set in a fibrous stroma. The cells may show nuclear pleomorphism.
The cells of the tumours stain black to brownish black with chromic acid, like some of the adrenal medullary tumours, but do not produce catecholamines. (Recently it has been shown that normal carotid body and its tumour sometimes contain considerable amounts of noradrenaline — W. Boyd.)
6. *The tumour is radio-resistant.*
8. Shape: Initially spherical but becomes irregular as it grows.
9. Surface: Usually smooth but may be bosselated. The edge is well defined.
10. *Consistence: Solid swelling*, firm to hard in feel. The tumour is often called as “potato tumour” because of its consistence, shape and size.
11. The tumour is not tender or hot. The overlying skin looks normal.
12. *Sometimes the tumour may pulsate.* This is either a transmitted pulsation from the adjacent carotid arteries, or a palpable external carotid artery which is pushed forward. Rarely, the tumour is itself a vascular one causing pulsation.
13. The lump is mobile from side to side, but not up and down.
14. The lump is deep to deep cervical fascia and lies beneath the anterior edge of sternomastoid muscle.

Diagnostic criteria

1. Age: Common in middle-aged individual (between 40 and 60 years).
2. Sex: Equally common in both sexes.
3. *The patient presents with a painless slowly growing lump in the upper anterolateral part of the neck.* The patient may notice that the lump pulsates. The lump increases very slowly and usually there is a long history.
4. He may also suffer from occasional fainting attack or blackout (transient cerebral ischaemia). This symptom is due to impaired cerebral circulation following compression of the internal carotid artery by the tumour at a certain posture. On application of pressure on the lump this syncopal attack may be produced.

Examination:

5. The lump is mostly unilateral.
6. *Site:* The lump is situated in the upper and lateral parts of the anterior triangle of the neck, beneath the anterior edge of sternomastoid muscle at the level of the upper border of thyroid cartilage (level of bifurcation of the common carotid artery).
7. *Size:* It varies but rarely exceeds the size of potato.

Differential diagnosis

1. Solitary metastatic neck lymph gland.
2. Koch's lymphadenitis.
3. Aneurysm of carotid artery.
4. Sternomastoid tumour.
5. Solid swelling in relation to the apex of the thyroid lobe.
6. Branchial cyst.

Complications

1. *Transient cerebral ischaemia leading to fainting attack* — due to impaired cerebral circulation following compression of the internal carotid artery by the tumour at a certain posture.
2. Dysphagia.
3. Change of voice following interference with the function of vocal cords due to pressure on the vagus nerve — rare.

Investigation

Carotid angiogram shows the displacement or splaying of the carotid fork. There may be presence of a mass of abnormal tumour vessels. *Angiogram also differentiates it from aneurysm of carotid artery.*

Treatment

The tumour is *radio-resistant*. So, *surgical removal is the treatment of choice*. Total excision is the most

successful form of treatment followed by partial excision and radiotherapy.

In some cases, the tumour can be separated from the fork of the carotid artery without injuring it by blunt and sharp dissection, and so removed.

In other cases, the tumour is large enough and inseparable from the fork of the carotid artery. So, the tumour is removed along with resection of arteries followed by a graft, e.g., autogenous vein — between common and internal carotid arteries to maintain the blood supply to the brain. This requires the use of hypothermia or some form of temporary bypass.

Danger during operation

Injury to the internal carotid artery—impairment of blood supply to the brain leads to contralateral hemiplegia.

Cautions

1. A general surgeon meeting unexpectedly with a carotid body tumour is best advised to confine himself to biopsy of the tumour. Most tumours may bleed profusely on biopsy.
2. In case of the old and enfeebled individuals with a long history of the lump the tumour may be left as it is without any interference.

STERNOMASTOID TUMOUR AND CONGENITAL WRY NECK (TORTICOLLIS)

This is a deformity characterised by tilting of the head towards the shoulder along with torsion of the neck and deviation of the head to the opposite side. In about 20 percent of cases, to start with, there is a hard lump over the sternomastoid noticed between 1 and 4 weeks; it disappears within a few months.

Causes

There are many theories to explain the development of this lesion:

1. *Ischaemia*: Probably there is an interference with the blood supply of the sternomastoid muscle, caused by injury during birth. It is held that the middle part of the sternomastoid muscle is supplied by an 'end artery', sternomastoid branch of the superior thyroid artery, and in wry neck the lumen of the sternomastoid artery has been found obliterated at the anterior border of

the muscle. It is suspected that the sternomastoid arteries might be obstructed during difficult labour, with subsequent fibrosis.

2. *Trauma*: The sternomastoid muscle might conceivably be torn during birth with the formation of haematoma. In the early months of life this haematoma appears as a lump (*the sternomastoid tumour of infancy*); but the lump is too discrete to be a haematoma, and microscopically contains no haemosiderin. The haematoma subsequently undergoes fibrosis followed by contracture.
3. *Congenital*: Hereditary theory is suspected by some authors, because such deformity may also be associated with other congenital deformities such as club-foot, congenital dislocation of hip, etc.
4. *Other theories*: Venous thrombosis or infective myositis has been suspected, but there is little convincing evidence.

Diagnostic criteria

1. During the first few weeks of life, the mother notices an elongated swelling in the middle of the sternomastoid muscle.
2. The swelling is hard in feel and tender, and the child cries bitterly when it is palpated, or if the muscle is stretched in any way.
3. Shape and size: The lump is fusiform with its long axis along the line of sternomastoid muscle. It varies in size from 1 to 2 cm across.
4. Surface and edge: The anterior and posterior edges of the lump are well defined but the inferior and superior edges are ill-defined.
5. Gradually the swelling and tenderness subside within a few months and the muscle becomes tense and contracted with the development of characteristic attitude. On examination at this stage the contracted muscle is felt as a tight cord.
6. Examine the spine gently. Neck movements are usually normal. There may be restriction of movements due to spasm of the sternomastoid muscle.
7. Examine the eyes to exclude squint. The torticollis may be secondary to squint.

Secondary changes

1. If the deformity is not corrected early, asymmetry of face develops due to adaptive

shortening of the other soft parts on the affected side. There are thickening and contraction of the deep cervical fascia, scalenus anterior muscle etc. The vessels also become shortened.

2. There may be secondary scoliotic deformity of the cervical and upper thoracic spines.

X-ray: Every case should be X-rayed to exclude any vertebral anomaly (e.g., hemivertebrae) which may be the primary error.

Treatment

1. *Prophylaxis:* If the sternomastoid tumour is detected immediately after birth, every effort should be made to prevent torticollis from developing. The neck should be manipulated daily into a position which elongates the affected sternomastoid to the full. The baby is laid on the bed to sleep on alternate sides.
2. *When the condition is detected only after the torticollis has been developed:*
 - (a) Daily physiotherapy to stretch the muscle and use of corrective brace — torticollis harness may be tried, but usually there is recurrence.
 - (b) **Operation:**
 - *Subcutaneous tenotomy at the lower end* — in an early and mild case, and where only the sternal head appears to be contracted. The danger is injury to blood vessels particularly internal jugular vein deep to the sternomastoid. The advantage is only for cosmetic reasons as the operation leaves no visible scar.
 - *Open division of both the heads of sternomastoid at the lower end.* Along with it, the deep cervical fascia, and if necessary the carotid sheath and scalenus anterior muscle, are also divided. The phrenic nerve and the internal jugular vein should be identified and safeguarded.
 - *Open division of the upper end of the sternomastoid* — may be performed alone, or may be combined with the above procedure. The spinal accessory nerve should be safeguarded. The advantage is that the scar is hidden by the hair.

After-treatment

After operation, the correction must be maintained by torticollis harness which should be worn for about 6

months. Physiotherapy — both active and passive movements of the neck should be continued for 6 months.

COLD ABSCESS IN THE NECK

On many occasions, a soft swelling, cold abscess, due to accumulation of pus of tuberculous origin, may be encountered in the region of the neck, commonly in the posterior triangle.

Why it is called cold abscess?

Because:

- there is no sign of inflammation and so the abscess is not hot and red as in pyogenic abscess;
- the abscess is a low tension abscess, so it is much less painful than pyogenic abscess.

Source of cold abscess in the neck

It may be:

1. Due to caseation of tuberculous lymph adenitis in the cervical region (Koch's lymphadenitis) — commonly found in the upper half of the anterior triangle of the neck.
2. Secondary to tuberculosis of the cervical spine (caries spine) — commonly found in the posterior triangle of the neck.

1. From Koch's lymphadenitis

Usually one group of cervical lymph nodes is infected, most frequently the upper jugular group of nodes is affected. The organism is tubercle bacilli, mostly 'human type'. Tubercle bacilli most commonly reach a lymph node by lymphatics, when the tubercles first form in the cortex. Blood-borne infection sometimes occurs, in which case the medulla of the lymph node is the first to be affected. The tubercle bacilli usually gain entrance through the tonsil of the corresponding side. The infective process passes through 3 stages:

1st stage: It is the stage of lymphadenitis. In this stage the lymph nodes become enlarged and slightly tender but remain discrete. If the resistance of the patient is good, the nodes may resolve with fibrosis and calcification.

2nd stage: In this stage, the lymph nodes become matted with perilymphadenitis.

3rd stage: In this stage, the caseating materials liquefy and break through the capsules of the lymph nodes to form a 'cold abscess'. In the beginning the

abscess remains deep to the deep cervical fascia. Ultimately the fascia is eroded at one point and the pus tracks into the superficial space. Now it is called '*collar-stud abscess*'. The superficial abscess gradually enlarges and becomes obvious on inspection. In untreated cases, the overlying skin becomes ischaemic and reddish blue at the centre of the abscess. Ultimately the skin at the centre necroses and gives way leading to a chronic discharging sinus.

2. From caries spine of the cervical region

The tuberculous process of the cervical spine may break through the bone and give rise to formation of cold abscess.

It may rupture anteriorly or posteriorly:

(a) *When it ruptures anteriorly* the pus will appear anteriorly first deep to the prevertebral layer of the deep cervical fascia from where the pus may travel in one of the following ways:

- In the upper cervical region the pus may pass forward and bulge anteriorly into the posterior wall of the pharynx when it will cause a chronic midline deep-seated swelling (cf. *Retropharyngeal abscess* which will cause an acute superficial swelling situated on one side of the midline).
- In the lower cervical region the pus may press the oesophagus and trachea forward.
- The pus may track downward behind the prevertebral fascia and enter the superior mediastinum.
- The pus may track laterally deep to the prevertebral fascia and behind the carotid sheath to

reach ultimately the posterior triangle beyond the posterior border of sternomastoid and thus form the cold abscess in the posterior triangle of the neck (Fig. 3.25).

(b) *When it ruptures posteriorly*, the pus enters the spinal canal. From here the pus may follow the anterior primary division of the cervical spinal nerves and thus appear in the posterior triangle of the neck (Fig. 3.25).

From the posterior triangle the pus can travel into the axilla along the axillary sheath which is the prolongation of prevertebral fascia through the apex of the axilla dragged down by the cords of the brachial plexus and the subclavian artery. From the axilla it may travel further down along the course of the brachial artery.

Diagnostic criteria

1. Age: Common in children, young adults, and in the elderly.
2. Sex: Equally common in both sexes.
3. The patient presents with a *swelling in the neck which gradually increases in size*. It is usually *painless*.
4. The patient may give history of previous solid neck swelling (i.e. enlarged neck lymph nodes) on which the area of softening has occurred.
5. Features of tuberculous toxæmia almost always present, i.e., evening rise of temperature, sweating, anorexia, loss of weight.
6. Family history of tuberculosis may be present.

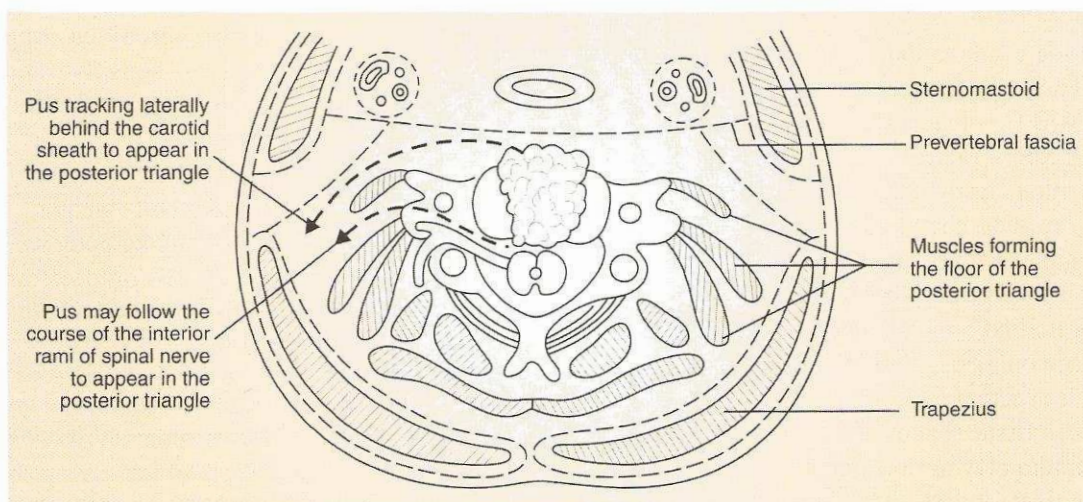


Fig. 3.25. The spreading of pus from the vertebral body to the posterior triangle.

Examination:

7. *Site*: The lump is commonly found in the upper half of the anterior triangle of the neck. It may be found in the posterior triangle.
8. *Size and shape*: Varies.
9. *Surface*: Irregular and indistinct. The overlying skin may be discoloured, *but there is no sign of inflammation* (particularly brawny induration of pyogenic abscess) unless secondarily infected.
10. *Edge*: Usually indistinct unless the abscess is tense.
11. *Consistence*: *A soft cystic swelling. Fluctuation is positive.*
12. *Usually not tender.*
13. A peculiar rim of thickening surrounding the central soft area may be present.
14. *Matted lymph nodes* may be palpable on the deeper aspect when the abscess is secondary to tuberculous lymphadenitis.
15. *Transillumination—negative* (cf. cystic hygroma).
16. *The cervical spine should always be examined to find:*
 - tenderness in the cervical spine;
 - limited movements of the neck;
 - rigidity of the spinal muscles.
17. General examination should be performed to see evidence of tuberculosis elsewhere, e.g., other lymph nodes, the lungs and the urinary tract.
18. *Aspiration of the abscess will show caseous material with no demonstrable cholesterol crystals* (cf. branchial cyst).

Investigations

1. Blood examination:
 - Hb% – anaemia.
 - W.B.C. – leucocytosis with lymphocytosis.
 - E.S.R. – raised.
2. Mantoux test – positive.
3. X-ray of the chest may show evidence of primary complex with enlarged mediastinal lymph nodes.
4. X-ray of the neck: A.P. & lateral views may show:
 - calcified lymph nodes;
 - osteoporosis, joint space diminution, areas of destruction in the cervical vertebrae;
 - soft tissue shadow, i.e., paravertebral shadow.
5. Sputum may or may not show acid-fast bacilli.
6. Aspiration of the abscess material and examination for *Mycobacterium tuberculosis*:

- Staining: Ziehl–Nielsen stain for acid-fast bacilli. Gram stain to exclude secondary infection.
- Culture in Lowenstein–Jensen media.
- Guinea pig inoculation test.

All the examinations will show positive results.

7. Lymph node biopsy, when the surrounding lymph nodes are palpable, confirms the diagnosis showing central area of caseation, surrounded by lymphocytes and giant cells.

Differential diagnosis

1. Branchial cyst: Here, the aspiration shows the cheesy material which contains cholesterol crystals.
2. Cystic hygroma: Here transillumination is brilliantly positive.

Treatment

1. General treatment, and
2. Treatment of the local condition.

General treatment

1. Anti-Koch's therapy: Anti-tuberculous drugs are to be used routinely and should be continued for a period of 18 months.
 - Inj. Streptomycin—0.75–1 gm daily for a total of 90–120 injections.
 - I.N.H. (Isonicotinic acid hydrazide)—300–400 mg daily.
 - P.A.S. (Para-amino-salicylic acid) – 10 gm daily.

The usual plan is to start with three drugs at the same time.

(Presently newer antituberculous drugs are available which are most sensitive and thus shorten the duration of treatment from 18 months to 9 months. These are:

- Rifampicin – 15 mg/kg body weight
 - Ethambutol – 25 mg/kg body weight
 - Pyrazinamide – 20–30 mg/kg body weight
2. Vitamins particularly B-complex and C. Minerals – iron preparation. Good food and healthy surroundings.

Local treatment

Includes treatment of the abscess and treatment of the primary source, if any.

- A. *Treatment of the abscess*: Usually the small abscess subsides with the general conservative

Examination:

7. *Site:* The lump is commonly found in the upper half of the anterior triangle of the neck. It may be found in the posterior triangle.
8. *Size and shape:* Varies.
9. *Surface:* Irregular and indistinct. The overlying skin may be discoloured, *but there is no sign of inflammation* (particularly brawny induration of pyogenic abscess) unless secondarily infected.
10. *Edge:* Usually indistinct unless the abscess is tense.
11. *Consistence:* *A soft cystic swelling. Fluctuation is positive.*
12. *Usually not tender.*
13. A peculiar rim of thickening surrounding the central soft area may be present.
14. *Matted lymph nodes* may be palpable on the deeper aspect when the abscess is secondary to tuberculous lymphadenitis.
15. *Transillumination—negative* (cf. cystic hygroma).
16. *The cervical spine should always be examined to find:*
 - tenderness in the cervical spine;
 - limited movements of the neck;
 - rigidity of the spinal muscles.
17. General examination should be performed to see evidence of tuberculosis elsewhere, e.g., other lymph nodes, the lungs and the urinary tract.
18. *Aspiration of the abscess will show caseous material with no demonstrable cholesterol crystals* (cf. branchial cyst).

Investigations

1. Blood examination:
 - Hb% – anaemia.
 - W.B.C. – leucocytosis with lymphocytosis.
 - E.S.R. – raised.
2. Mantoux test – positive.
3. X-ray of the chest may show evidence of primary complex with enlarged mediastinal lymph nodes.
4. X-ray of the neck: A.P. & lateral views may show:
 - calcified lymph nodes;
 - osteoporosis, joint space diminution, areas of destruction in the cervical vertebrae;
 - soft tissue shadow, i.e., paravertebral shadow.
5. Sputum may or may not show acid-fast bacilli.
6. Aspiration of the abscess material and examination for *Mycobacterium tuberculosis*:

- Staining: Ziehl–Nielsen stain for acid-fast bacilli. Gram stain to exclude secondary infection.
- Culture in Lowenstein–Jensen media.
- Guinea pig inoculation test.

All the examinations will show positive results.

7. Lymph node biopsy, when the surrounding lymph nodes are palpable, confirms the diagnosis showing central area of caseation, surrounded by lymphocytes and giant cells.

Differential diagnosis

1. Branchial cyst: Here, the aspiration shows the cheesy material which contains cholesterol crystals.
2. Cystic hygroma: Here transillumination is brilliantly positive.

Treatment

1. General treatment, and
2. Treatment of the local condition.

General treatment

1. Anti-Koch's therapy: Anti-tuberculous drugs are to be used routinely and should be continued for a period of 18 months.
 - Inj. Streptomycin—0.75–1 gm daily for a total of 90–120 injections.
 - I.N.H. (Isonicotinic acid hydrazide) – 300–400 mg daily.
 - P.A.S. (Para-amino-salicylic acid) – 10 gm daily.

The usual plan is to start with three drugs at the same time.

(Presently newer antituberculous drugs are available which are most sensitive and thus shorten the duration of treatment from 18 months to 9 months. These are:

- Rifampicin – 15 mg/kg body weight
 - Ethambutol – 25 mg/kg body weight
 - Pyrazinamide – 20–30 mg/kg body weight
2. Vitamins particularly B-complex and C. Minerals – iron preparation. Good food and healthy surroundings.

Local treatment

Includes treatment of the abscess and treatment of the primary source, if any.

- A. *Treatment of the abscess:* Usually the small abscess subsides with the general conservative

treatment. If the abscess is a bigger one then aspiration, in addition, is helpful.

Techniques of aspiration: The needle of aspiration should be insinuated through the healthy skin; otherwise, there is a chance of sinus formation if the damaged skin is punctured. The aspiration should be done from a higher level and never at dependent part. Otherwise, there is a chance of sinus formation along the track of the needle.

- B. If there is associated glandular enlargement not responding to conservative treatment — excision biopsy of the tubercular lymph glands as a whole may be done.
- C. *When the lesion is secondary to Koch's vertebrae:*
 1. Immobilisation of the cervical spine with the help of plaster jacket (Minerva jacket).

Extent of Minerva jacket:

- Below: Middle of the chest or nipple line.
- Above: The plaster extends above to cover the neck and scalp exposing the ear and face.

The plaster immobilisation is continued for 3 to 6 months.

2. **Operation:** Cervical spine fusion. It may be done when the plaster immobilisation fails, or in the beginning.

COLLAR-STUD ABSCESS

It is a bilocular abscess with an intervening narrow communication (Fig. 3.26).

Causes

1. **Pyogenic:** Commonly found in the palm. The superficial loculus lies superficial to the palmar fascia and the deep loculus is situated deep to the palmar fascia with a narrow communication connecting the two. The communication occurs either due to erosion of the fascia or due to rupture of the fascia by increased tension.
2. **Tuberculous:** Commonly found in the neck, due to caseation of tubercular lymph node followed by formation of cold abscess. For some time the abscess remains limited by the resistance of the deep cervical fascia. Ultimately the deep fascia is eroded at one point and the pus tracks into the superficial space.

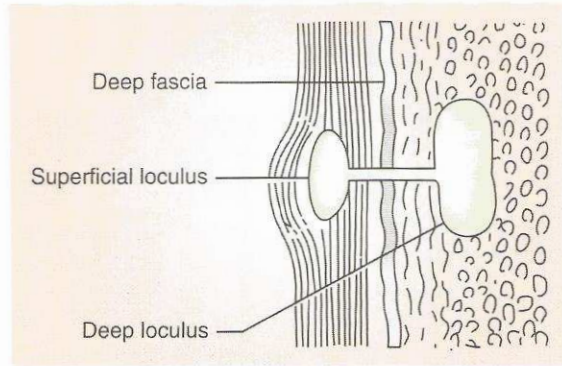


Fig. 3.26. The superficial and deep loculi of a collar-stud abscess.

Causes of non-tuberculous cold abscess

1. Leprosy: Nodular leprosy with degeneration.
2. Actinomycosis: Abscess following actinomycotic infection.
3. Gumma degeneration.

Differential diagnosis of neck swellings

While examining a neck swelling one should first observe *where the lump is situated* — *anterior triangle or posterior triangle*. All midline swellings are included within the anterior triangle. If the lump is situated in the anterior triangle observe *whether the lump moves on swallowing*. If it moves on swallowing observe *whether the lump moves up on protrusion of the tongue*. Then one should examine the lump for other clinical characteristics such as single or multiple, solid or cystic, transilluminable or not, pulsatile or non-pulsatile. This protocol helps to define a lump clinically in most of the cases. Multiple lumps are invariably lymph nodes. *One should note that the commonest lump in the neck is of lymph node origin.*

One should remember also that any lump arising from skin and subcutaneous tissue such as, lipoma, fibroma, neurofibroma, sebaceous cyst, and haemangioma can occur anywhere in the neck.

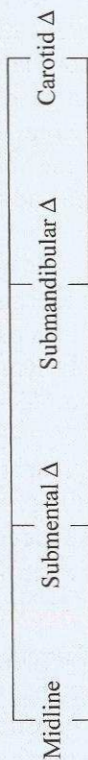
Pulsatile swellings in the neck

1. Aneurysm of the carotid artery.
2. Aneurysm of the subclavian artery.
3. Carotid body tumour — transmitted pulsation.
4. Lymph node swelling in relation to the carotid artery — transmitted pulsation.
5. Primary toxic goitre — occasionally.

Algorithm for Examination of Neck Swelling

SWELLING IN THE NECK

Site

Anterior triangle (Δ)

Observe movements on swallowing

Moves

Observe movements on protrusion of the tongue

Does not move

Does not move

Solid

1. Thyroid gland
2. Ectopic thyroid
3. Pretracheal lymph node

Cystic

1. Subhyoid bursal cyst
2. Cyst in relation to the thyroid isthmus
3. Laryngocele

Moves

Thyroglossal cyst

Midline

Solid

1. Submental lymph node
2. Bony growth arising from manubrium

Cystic

1. Ranula
 - Transillumination—positive
2. Cervical dermoid
 - Transillumination—negative
3. Cold abscess in the space of burns
 - Transillumination—negative
 - Aspiration—cheesy pus
4. Innominate aneurysm
 - Pulsatile

Submental Δ (see Midline)**Solid**

Lymph node

Cystic

1. Ranula
2. Cervical dermoid

Submandibular Δ **Solid**

1. Submandibular salivary gland
 - Bimanually palpable
2. Lymph node

Cystic

1. Plunging ranula
 - Transillumination—positive
2. Lateral variety of sublingual dermoid
 - Transillumination—negative

Carotid Δ **Solid**

1. Lymph node
2. Carotid body tumour

Cystic

1. Branchial cyst
 - Aspiration—cholesterol crystals
2. Cold abscess
 - Aspiration—cheesy pus
3. Carotid aneurysm
 - Pulsatile

Posterior triangle (Δ)**Solid**

1. Lymph node
2. Cervical rib

Cystic

1. Cystic hygroma
 - Transillumination—positive
2. Cold abscess
 - Transillumination—negative
 - Aspiration—cheesy pus
3. Pharyngeal pouch
4. Subclavian aneurysm
 - Pulsatile

4

CHAPTER



Cervical Lymphadenopathy

DISTRIBUTION OF CERVICAL (NECK) LYMPH NODES

Lymph nodes in the neck are arranged in two groups:

- I. Superficial group: These are a few and are scattered superficial to the investing layer of deep cervical fascia.
- II. Deep group: These are situated deep to the investing layer of deep cervical fascia and are distributed in two groups:
 1. Vertical group
 2. Circular group

Vertical group

Lying in relation to the internal jugular vein and deep to the sternomastoid muscle.

1. *Jugulodigastric nodes* — lying below the posterior belly of digastric muscle as it crosses the internal jugular vein. It also includes the tonsillar lymph node.
2. *Juguloomohyoid nodes* — lying behind the internal jugular vein where it is crossed by the anterior belly of the omohyoid muscle. All the lymph from the tongue ultimately reaches this group.
3. *Supraclavicular nodes* — lying around the inferior part of the internal jugular vein behind the sternomastoid muscle extending into supraclavicular region.

Virchow's glands

These are left supraclavicular group of lymph glands lying *between the two heads of sternomastoid*. These are the common sites of metastases from the gastric carcinoma, testicular tumour, oesophageal carcinoma, and when involved, the prognosis is grave. Enlargement of these nodes with abdominal malignant growths is known as *Troisier's sign*.

Circular group

From anterior to posterior:

1. Submental
2. Submandibular
3. Pre-auricular
4. Post-auricular
5. Occipital.

The cervical groups of lymph nodes can also be described as **six levels of lymph nodes from level I to level VI to facilitate lymph node dissection.**

- Level I** Submental (IA) and submandibular (IB) lymph nodes.
- Level II** Lymph nodes in relation to upper third of internal jugular vein — jugulodigastric nodes.
- Level III** Lymph nodes in relation to middle third of internal jugular vein — juguloomohyoid nodes.
- Level IV** Lymph nodes in relation to lower third of internal jugular vein.
- Level V** Lymph nodes in the posterior triangle of the neck. Supraclavicular nodes are included in this group.
- Level VI** Pre and paratracheal nodes.

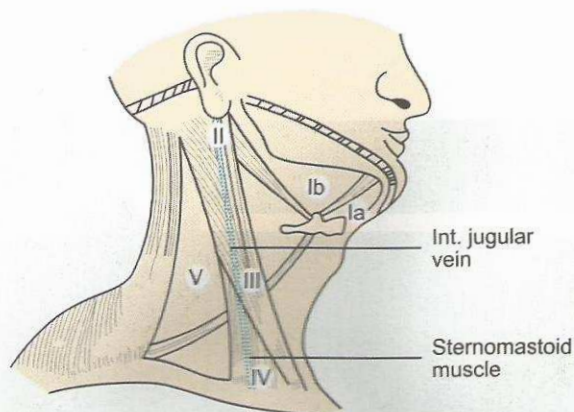


Fig. 4.1. Different levels of cervical lymph nodes.

EXAMINATION OF ENLARGED LYMPH NODE

Patients presenting with enlarged lymph node or nodes are a common clinical problem. They usually present with a lump in any particular area, e.g., neck or groin. Lymph nodes swellings can have different consistencies depending on the underlying pathology.

Consistence of the lymph nodes:

- *Firm* in cases of reactive hyperplasia and tuberculosis.
- *Rubbery* in cases of Hodgkin's lymphoma.
- *Hard* in secondary neoplasms.

When one suspects that the lump is of lymph node origin, one must complete the examination by fulfilling the following five requirements in addition to an examination of the lump proper.

1. *Examine the drainage area of the lymph nodes for any possible primary lesion, either infection or malignancy, e.g.*
 - for enlarged lymph nodes in the neck – examine the scalp, ear, nose, mouth cavity and tongue, larynx and pharynx, arms and breast;
 - for enlarged lymph nodes in the groin – examine the skin of the leg, buttock and lower abdominal wall below the umbilicus and the external genitalia and anus.
2. *Examine the other lymph node areas, e.g., contralateral neck, axilla, groin, abdomen particularly where no primary source has been detected.*

In generalised lymphadenopathy enlarged lymph nodes are also found at other sites.

3. *Examine the abdomen for:*

- **Liver and spleen:** If enlarged, suggest a **reticulosis, sarcoid** or glandular fever.

- **Any mass:**
 - the mass may be enlarged retroperitoneal nodes, e.g., in seminoma testis.
 - the mass may be primary carcinoma of any intra-abdominal viscus, e.g., cancer stomach or pancreas (cf. Virchow's gland).

4. *Examine the testis, e.g., testicular tumour.*
5. *Rectal and vaginal examination* – to exclude any rectal or prostatic growth, or ovarian tumour.

Investigations

1. **Blood:**
 - Routine examination – Hb%, WBC count, ESR may give clues to infection, malignancy or reticuloses.
 - Examination of blood film – may give a clue to leukaemia or glandular fever.
 - Examination of blood for W.R. and Kahn – if syphilis is suspected.
2. **Chest X-ray** – To note:
 - A primary lesion of the lung
 - Koch's
 - Tumour
 - Evidence of any secondary metastasis in the lung.
 - Evidence of enlarged mediastinal nodes.
3. **X-ray of the neck** – enlarged painless cervical nodes may show typical spotty calcification in tuberculous nodes. Further investigations will depend on the clinical findings and the routine investigations.
4. *Biopsy of one enlarged lymph node* – may be necessary for a definite histological proof of the diagnosis, e.g., tuberculosis, Hodgkin's disease, lymphosarcoma, or for knowing the nature of a hidden primary lesion.
5. *Biopsy of any visible primary lesion in the drainage area of the lymph nodes* should be performed before planning an attack on the enlarged node or nodes.
6. **Laryngoscopy** – In the presence of enlarged hard nodes in the neck, when no visible primary lesion is evident, one should perform laryngoscopy to exclude any growth in the posterior one-third of the tongue, larynx and pharynx. Posterior third growth of the tongue usually presents with enlarged neck nodes.

CERVICAL LYMPHADENOPATHY

Enlargement of the cervical lymph node (or nodes) is by far the commonest swelling in the neck.

Causes

Acute

1. Acute pyogenic lymphadenitis
2. Acute lymphatic leukaemia
3. Acute infectious mononucleosis – caused by E.B. virus.

Chronic

I. Inflammatory

1. Non-specific: Chronic pyogenic lymphadenitis
2. Specific:
 - Tuberculosis
 - Syphilis – in secondary stage
 - Sarcoidosis
 - Brucellosis
 - Fungal diseases like blastomycosis.

II. Neoplastic

Almost always malignant.

1. Primary:
 - Lymphoma – Hodgkin's disease
 - Lymphosarcoma
 - Reticulum cell sarcoma
 - Chronic lymphatic leukaemia
 - Burkitt's lymphoma.
2. Secondary: Metastatic lymph node from a primary growth, mostly from carcinoma or melanoma.

III. Autoimmune disorders

1. Systemic lupus erythematosus
2. Juvenile rheumatoid arthritis (Still's disease).

Causes of generalised lymphadenopathy

I. Inflammatory

- Acute
 - Infectious mononucleosis
 - Septicaemia
- Chronic
 - Tuberculosis
 - Syphilis in secondary stage.

II. Reticuloses

- Hodgkin's disease
- Lymphosarcoma
- Lymphatic leukaemia

III. Sarcoidosis

Differential diagnosis of chronic cervical lymphadenopathy

I. Chronic pyogenic lymphadenitis

1. *Lymph nodes: Firm and tender but not matted.*
2. Chronic infection in the drainage area particularly in the oral cavity.

3. Blood examination: Leucocytosis with increased polymorphs.
4. Lymph node biopsy shows sinus hyperplasia.

II. Tuberculous lymphadenitis

1. Common in children, but may occur at any age.
2. *Painless enlargement of the lymph nodes.*
3. Features of tuberculous toxæmia: Mild degree fever with evening rise, anorexia, anaemia, loss of weight, cough, etc.
4. Characteristics of enlargement:
 - Cervical group is enlarged earliest.
 - Occasionally the enlargement may be generalised.
 - The enlarged glands are firm in feel and initially discrete but *later matted* (due to *peradenitis*) and often slightly tender. There is no sign of inflammation unless secondarily infected. In the late stage, there may be *cold abscess* formation due to caseation and liquefaction, or there may be *sinus* or *ulcer formation* over the enlarged nodes which refuses to heal (Figs. 4.2 and 4.3).
 - Mediastinal group may be enlarged in children and, therefore, may cause features of mediastinal compression.
5. Evidence of tuberculosis in the lung may be present.
6. Investigations:
 - Blood examination: Anaemia, leucocytosis with lymphocytosis and high E.S.R.
 - Mantoux test: Positive.
 - Chest X-ray: May show evidence of primary complex with enlarged mediastinal lymph node.
 - X-ray of the neck: May show calcified lymph node.
 - Sputum may or may not show acid-fast bacilli.
 - *Biopsy of the lymph node:* Confirms the diagnosis by showing the central area of caseation, surrounded by epithelioid cells, lymphocytes and giant cells.



Fig. 4.2. Koch's ulcer with lymphadenitis.



Fig. 4.3. Tubercular sinus left supraclavicular region. Also note the adjacent lymph node swelling.

III. Secondary metastatic lymph node

1. **Age:** Common in elderly individuals since cancers mostly occur in patients over the age of 50. The exception is papillary carcinoma of thyroid which occurs in adolescents and young adults. Metastatic cervical nodes in children are usually due to primary thyroid carcinoma or neuroblastoma.
2. **Sex:** Mostly common in men.
3. The patient presents with a *painless enlarged lump in the neck*. The lump usually grows slowly and new crops of lump may appear.
4. General symptoms of anorexia, weight loss, weakness may be present, but these are usually late and rare with head and neck cancers.
5. The patient may have symptoms of the primary lesion such as ulcer in the tongue, sore throat, hoarseness of voice, cough, haemoptysis, dysphagia, etc.

Examination:

6. **Site:** Metastatic nodes are more common in the nodes of the anterior triangle. These are deep to the anterior edge of the sternomastoid muscle.
7. **Characteristics of enlarged lymph nodes:** The enlarged nodes are stony hard, mobile or fixed, not tender. In the early stage, the nodes are smooth and discrete and of variable sizes. As they grow they may coalesce into one large mass with irregular or bosselated surface. The overlying skin usually looks normal unless infiltrated.

8. Evidence of primary growth is almost always present commonly in one of the following sites:
 - (a) Scalp, face, neck (e.g., thyroid, parotid), ear.
 - (b) Buccal cavity – Lips, tongue, buccal mucous membrane, tonsils.
 - (c) Nasal cavity and maxillary antrum.
 - (d) Pharynx, larynx – *So, examine with mirrors.*
 - (e) Upper limb and chest wall.
 - (f) Thorax – The lungs, oesophagus.
 - (g) Breast.
 - (h) Abdomen – The stomach, pancreas, ovaries.
 - (i) External genitalia – The testes.
 (*So, the above mentioned sites should be examined in each case to detect the site of the primary lesion. Nature of the primary tumour is commonly either carcinoma or melanoma.*)

9. Presence of enlarged metastatic lymph nodes in the left supraclavicular fossa lying between two heads of sternomastoid muscle is known as *Virchow's gland*. Its presence in association with abdominal malignant growths is called *Troisier's sign*.
10. Investigation – Apart from routine investigations the nature of the special investigations will be mostly dictated by the primary site of the lesion.
11. Biopsy of the enlarged lymph nodes occasionally may be helpful in detecting the hidden primary lesion.

IV. Hodgkin's disease

1. **Age:** It may occur at any age but is common in the young between 15 and 30 years and in the old age above 40 years.
2. **Sex:** Common in males.
3. The patient usually presents with *slowly growing, discrete, painless, rubbery enlargement of accessible nodes commonly in the neck*. The superficial groups of cervical lymph nodes are enlarged first which then increase progressively to involve the other groups to become generalised in nature.
4. Presence of constitutional symptoms as follows:
 - (a) Fever with or without rigors occurring in a periodic fashion (i.e., a period of pyrexia for 5 to 10 days alternating with nearly a similar period of apyrexia which goes on for several months – *Pel-Ebstein fever*. Rarely, in advanced cases the temperature may be nearly continuous.

- (b) Weight loss.
- (c) Night sweats.
- (d) Anaemia or pallor.
- (e) Itching of the skin (pruritus) – an unexplained but distinctive complaint.
- (f) Pain in the bones due to lymphomatous infiltration of the skeleton.
- (g) Pain in the abdomen.

These pains or itching may be induced or enhanced by drinking alcohol – a peculiar and unexplained feature.

5. There may be dyspnoea, hoarseness of voice with features of mediastinal compression syndrome like venous congestion in the neck with the development of collateral veins across the chest wall due to enlarged mediastinal nodes. Rarely, the patient may present with oedema of both legs due to obstructions of inferior vena cava with large mass in the abdomen.

Local examination:

6. *Site:* Any of the cervical lymph nodes can be affected but lymphoma often causes lymphadenopathy in the posterior triangle, commonly in the supraclavicular fossa.
7. *Characteristics of enlarged lymph nodes:* Usually asymmetrical, moderate to severe degrees of enlargement. The nodes are ovoid, smooth and discrete. *These are solid, firm and rubbery in consistence and are not tender.* Usually mobile, but rarely, completely fixed. Occasionally, the lymph nodes may be matted in late stages (pseudo-matting). *The surrounding structures and the overlying skin are usually normal* (cf. Koch's lymphadenitis).

8. General examination:

- The patient may be anaemic with peculiar pallor like white-coffee, and may be jaundiced.
- Other groups of lymph nodes may be enlarged. *(So, palpate the lymph nodes in other areas of the body as well, e.g. axilla, abdomen and groin.)*
- Moderate degree enlargement of spleen and liver, with or without enlarged abdominal lymph nodes. *(So, palpate the abdomen in each case. Testes should also be palpated.)*
- Evidence of mediastinal compressions, pleural effusion may be present.
- Enlarged mediastinal nodes may occlude the

superior vena cava and cause venous congestion in the neck and the development of collateral veins across the chest wall.

- Oedema of the legs due to obstruction of the inferior vena cava by large masses in the abdomen.
- Rarely, there may be elevated, reddened, scaly patches of the skin, known as *mycosis fungoides* due to spread of tumour cells in the skin.

9. Investigations:

- Blood examination:
 - Anaemia
 - Pancytopenia
 - Leucocytosis with lymphopenia but eosinophilia.
- Chest X-ray—may show enlarged mediastinal shadow and pleural effusion.
- *Gordon's test*—Intracerebral injection of an emulsion of the human gland extract into the rabbit causes encephalitis.
- CT scan—may be helpful to detect involvement of the retroperitoneal and mediastinal lymph nodes, liver and spleen.
- Lymph node biopsy—confirms the diagnosis.
 - *Macroscopic examination:* Enlarged node without any evidence of periadenitis. *Cut section shows fish-flesh appearance.*
 - *Microscopic examination:* Central area of necrosis surrounded by various types of cells like eosinophils, polymorphs, lymphocytes, plasma cells, reticulum cells and characteristic giant cells popularly known as '*Reed-Sternberg's giant cells*'. *Cellular pleomorphism is the characteristic feature.*
- Exploratory laparotomy—may be helpful to detect involvement of retroperitoneal lymph nodes, liver and spleen.

V. Lymphosarcoma and reticulum cell sarcoma (non-Hodgkin's lymphoma)

The features are more or less the same as in Hodgkin's disease except that *prolonged duration is against lymphosarcoma and reticulum cell sarcoma.*

1. *Age:* Common in children or younger age group.
2. Presence of constitutional symptoms like loss of weight, anaemia, anorexia, gross weakness. Unexplained fever is present in 25 percent cases.

3. *Rapidly growing swelling in the neck.*
4. The swelling rapidly loses its normal ovoid outline.
5. The overlying skin is tense and shiny, and shows engorged veins.
6. Surface is irregular and consistence is heterogeneous, i.e., some are soft to firm and some are hard.
7. Chest X-ray – may show enlarged mediastinal nodes.
8. Lymph node biopsy – confirms the diagnosis.

VI. Chronic lymphatic leukaemia

1. *Age:* Usually *above 50 years*, commonly in males.
2. Presence of constitutional symptoms like fever, gross weakness, loss of weight, and recurrent upper respiratory tract infection.
3. There is painless, slowly growing progressive enlargement of the cervical lymph nodes and it ultimately becomes generalised.
4. *On examination:*
 - Anaemia – Moderate.
 - Lymph nodes – Either localised or generalised enlargement, discrete, firm in feel, mobile and not tender.
 - Skin – may show nodules or thickening of skin due to leukaemic tissue infiltration.
 - Spleen and liver – Moderately enlarged, firm, smooth and not tender.
5. *Investigations:*
 - Blood examination – W.B.C. count 80,000/mm³ to 1 lakh/mm³ with 90 percent lymphocytes.
 - Lymph node biopsy – Confirms the diagnosis and shows proliferation of the lymphocytes and the germinal centre.

VII. Syphilis in the secondary stage

1. *Age:* 20 to 25 years.
2. History of exposure: Present.
3. Symptoms: Ulcers in the mouth or genitalia, fever, arthritis and various skin rash.
4. *On examination:*
 - *Mucocutaneous lesion:* Ulcers on the dorsum of the tongue, angular fissure and condyloma.
 - Pleomorphic type of skin rash.
 - *Lymph nodes:* Generalised enlargement of the superficial group, mild degree, *firm in feel*,

discrete and shotty, not tender. Most characteristically there is *enlargement of epitrochlear and suboccipital groups.*

5. Investigation:

- W.R. and Kahn test: Strongly positive.
- *Treponema pallidum:* May be demonstrated in the exudate collected from the mucocutaneous lesion.

VIII. Sarcoidosis

1. *Age:* Common in young adults and middle-aged persons.
2. (a) Presence of constitutional symptoms which are extremely variable – fever, pain in the bones, paroxysmal dyspnoea due to bronchospasm, pain in the eyes with redness, dysphagia. May present with Raynaud's phenomenon.
- (b) May present with enlarged superficial groups of lymph nodes.
3. *On examination:*
 - Uveitis and conjunctivitis.
 - Enlargement of salivary gland, particularly the parotid gland.
 - Lymph nodes: Superficial groups are enlarged as a whole and most characteristically the preauricular groups. The qualities of the lymph nodes are like those of Hodgkin's, i.e., firm, discrete, not tender.
 - Facial nerve paralysis is the characteristic feature.
4. *Investigation:*
 - Blood examination – shows hyperglobulinaemia and hypercalcaemia.
 - X-ray of the phalanges – shows punched out appearance.
 - Chest X-ray – shows enlarged mediastinal lymph nodes.
 - *Kveim test* – positive and diagnostic.
 - Lymph node biopsy – may add further diagnostic information. Histology shows follicles of epithelioid cells and giant cells in the acute stage which may eventually progress to irreversible hyaline fibrosis.

LYMPHOMAS

Lymphomas are malignant neoplasms of the

lymp
splee
parts
lymph
of lym
deriva

Aetio

Exact
implic

1. V
2. In
3. D
4. Ic

Class

1. H
2. N
3. D
4. Ic

Organ

Nodal
Extran
Cells o

B ly
produ
T lym
kill th
work

HOD

The d
1982.
reticu

lymphoreticular system that includes lymph nodes, spleen, liver, thymus gland and bone marrow; all are parts of the lymphatic system. *They arise from the lymphocytes.* They are characterised by proliferation of lymphocytes, histiocytes and their precursors and derivatives.

Aetiology

Exact aetiology is not known. Following have been implicated:

1. Virus – Epstein-Barr (DNA) virus is associated with Burkitt's lymphoma.
2. Immunodeficiency diseases – inherited or acquired.
 - Associated with a primary immune disorder.
 - Associated with the human immunodeficiency virus (HIV).
 - Associated with post-transplant.
3. Drugs – Phenytoin induces lymphoma-like syndrome.
4. Ionising radiation.

Classification of malignant lymphoma

1. Hodgkin's lymphoma (HL) – characterised by cellular pleomorphism and the presence of Reed-Sternberg giant cells (RS cells).
2. Non-Hodgkin's lymphoma (NHL) – characterised by cellular pleomorphism *but RS cells are absent.*

Organ involved	HL	NHL
Nodal	90%	60%
Extranodal	10%	40%
Cells of origin	B lymphocytes	B lymphocytes, T lymphocytes, Null cell, Histiocytes (rare)

B lymphocytes become plasma cells which in turn produce antibodies that neutralise foreign invaders. T lymphocytes are derived from the thymus. T cells kill the foreign invaders directly. B cells normally work with T cells.

HODGKIN'S LYMPHOMA (HL)

The disease was described by Thomas Hodgkin in 182. This is malignant neoplasm of the lymphoreticular system and *arises from B lymphocytes.* This

is the commonest type of lymphoma and can occur wherever there is lymphoid tissue.

Pathology

- The disease *usually begins in a single lymph node* as a painless mass and then spreads to adjacent lymph nodes which are also enlarged but *without matting* (cf. Koch's lymphadenitis).
- Commonly involves the node(s) in the left supra-clavicular region.
- Spread then may occur to the other groups of lymph nodes in an orderly fashion i.e., progressing from one group of lymph nodes to next group.
- The axial lymphatic system is almost always affected i.e. mediastinal and para-aortic lymph nodes.
- Cut surfaces of the lymph nodes are smooth, homogenous and pink-grey in colour – *fish flesh appearance.*
- Microscopically, *cellular pleomorphism* is noted with lymphocytes, histiocytes, eosinophils and Reed-Sternberg cells (RS cells). *RS cells are the hallmark of Hodgkin's lymphoma.*

Reed-Sternberg cell – A giant cell derived from the abnormal B lymphocytes. It contains two large nuclei which are mirror images of each other – *looks like owl's eye.*

Presence of RS cells confirms the diagnosis of HL though absence of RS cells does not exclude the diagnosis. *RS cells are absent in NHL.*

The disease can involve the spleen, liver, bone marrow in the vertebral column and pelvis. Rarely, it may be confined to a single organ such as stomach.

New histological classification

- Rye classification is no longer used.
- REAL (Revised European American Lymphoma) classification is in use which divides HL in two groups:
 - (a) *Nodular lymphocyte predominant* – prognosis very good.
 - (b) *Classic Hodgkin's lymphoma*:
 - (i) *Nodular sclerosis* – most common type; prognosis good.
 - (ii) *Mixed cellularity* – prognosis fair.
 - (iii) *Lymphocyte depleted* – prognosis poor.
 - (iv) *Lymphocyte-rich* – a new entity.

Clinical features

1. *Age*: HL has a *bimodal age-specific* incidence rate; one mode occurring at ages between 15 and 30 years and the other mode above age 50.
HL in children under 10 years of age is seen more frequently in underdeveloped countries.
2. *Sex*: Common in males.
3. The patient usually presents with a *slowly growing, painless lump in the neck*, commonly supraclavicular region. The lump may be a single enlarged node or multiple but discrete enlarged nodes (*not matted*: cf. Koch's lymphadenitis).
4. The patient may present with generalised lymphadenopathy when more than one group of lymph nodes are enlarged e.g. neck, axillary, inguinal, mediastinal and para-aortic nodes are involved.
5. In addition to lymphadenopathy, the patient may present with *systemic symptoms* of fever, night sweats, weight loss, malaise, anaemia and pruritis – these are known as 'B symptoms'. *B symptoms indicate poor prognosis*.
 - (a) Fever with or without night sweats is *usually remittent*. Occasionally the fever occurs in a cyclical pattern called *Pel-Ebstein fever*, which is characterised by a period of pyrexia for 5 to 10 days alternating with nearly similar period of apyrexia which goes on for several months. Rarely, in advanced cases, the fever may be nearly continuous.
 - (b) Drenching night sweats.
 - (c) Weight loss more than 10% of body weight in the previous 6 months.
 - (d) Pruritis – itching of the skin. An unexplained but distinctive complaint due to heavy eosinophilic infiltration at the sites involved by the tumour.
 - (e) *Alcohol-induced pain or pruritis after drinking alcohol* – a peculiar feature of HL.
 - (f) Pain in the bones due to bone involvement which are usually osteoblastic/osteosclerotic. *Pathological fractures are rare*.
6. Patient may also present with symptoms due to pathological compression:
 - (i) Dyspnoea, hoarseness of voice, features of mediastinal compression like venous congestion and engorgement in the neck,

with the development of collateral veins across the chest wall due to superior venacaval obstruction by the enlarged mediastinal nodes.

- (ii) Rarely, the patient may present with oedema of the legs due to lymphatic and/or inferior venacaval obstruction by large masses of para-aortic nodes in the abdomen.

Examination**1. Local examination:**

- (a) The lump in the neck may be an enlarged single lymph node more than 2 cm in diameter.
- (b) The lump may be moderate to severe degrees enlarged lymph nodes with a "bunch of grapes" appearance.
The nodes are ovoid, smooth, solid, firm and rubbery and non-tender. Usually mobile, neither fixed to deeper structures nor to the overlying skin. The enlarged nodes remain discrete till the late stage when the nodes may be matted—*pseudomattening* (cf. Koch's lymphadenitis).
- (c) The surrounding structures and overlying skin are usually normal (cf. local signs of inflammation with or without abscess in septic lymphadenitis; cold abscess, sinus or ulcers in Koch's lymphadenitis).
- (d) The enlarged nodes are neither warm nor tender.

2. General examination:

- (a) Progressive anaemia with peculiar pallor—*coffee white pallor*.
- (b) Jaundice—due to excessive haemolysis of the red cells or involvement of liver or compression of CBD by enlarged glands at porta hepatis.
- (c) Enlargement of other groups of lymph nodes—*generalised lymphadenopathy*; so palpate for lymphadenopathy in other parts of the body e.g. opposite neck, axillae, groins and abdomen.
- (d) Enlargement of spleen, liver and para-aortic lymph nodes may be noted on abdominal palpation.
- (e) Venous congestion in the neck, engorged collateral veins across the chest wall due to mediastinal compression causing superior venacaval obstruction.
- (f) Presence of sternal and bony tenderness due to bone marrow involvement.

- ② Rarely, elevated, reddened scaly patches on the skin known as *mycosis fungoides* may be noted. It is not a fungal infection but is caused due to infiltration of the skin with malignant deposits.

Investigations

1. Complete blood count, ESR.
 - (i) Anaemia – normocytic normochromic.
 - (ii) Leukaemoid reaction.
 - (iii) Eosinophilia.
 - (iv) Lymphocytopenia – indicates bad prognosis.
 - (v) High ESR – indicates persistent disease activity.
2. Excision biopsy of the lymph node
 - (i) To establish diagnosis.
 - (ii) To do accurate histological grading.
 - (iii) To test the immune markers.

Before proceeding to any sophisticated investigations biopsy of the lymph node(s) is essential.

 - FNAC may give the diagnosis but fails to provide definite histological pattern.
 - Biopsy with a spring-loaded Trucut needle under image control can provide sufficient tissue for histological grading.
 - Excision biopsy is preferred whenever possible to provide both diagnosis and histological pattern.
 - CT-guided FNAC may be performed in abdominal lymphoma and thus to avoid laparotomy.
3. Chest X-ray: to look for
 - (i) Widening of mediastinum.
 - (ii) Pleural effusion.
 - (iii) Lung parenchymal infiltration.
4. US, CT, MRI of chest and abdomen: to look for
 - (i) Mediastinal lymph node enlargement.
 - (ii) Para-aortic lymph node enlargement.
 - (iii) Liver secondaries.
 - (iv) Splenomegaly.
5. PET scan: can detect small deposits as low as 0.2 cm, that do not show on CT scan.
6. IVU: to note any distortion or compression of the renal calyces or pelvis by the enlarged para-aortic lymph nodes – rarely performed.
7. Lymphangiography – rarely performed.

8. Gallium scan.
9. Other blood investigations
 - LFT, serum alkaline phosphatase – increased levels indicate liver involvement.
 - Urea, creatinine – increased levels indicate renal involvement.
 - Uric acid – increased levels indicate aggressive NHL.
 - HIV testing – high grade NHL may be associated with AIDS.
10. Skeletal survey and/or bone scan:
 - To include thoracic and lumbar vertebrae, pelvis, proximal limbs and any other bone which may be tender.
 - Usually *osteosclerotic lesion with HL* and *osteolytic lesion with NHL*.
11. Bone marrow aspiration or trephine biopsy in presence of
 - ↑ Serum alkaline phosphatase
 - Abnormal skeletal survey
 - Stage III disease
 - Unexplained anaemia
12. Staging laparotomy.

Staging laparotomy followed by splenectomy and liver and lymph node biopsy are not performed routinely now-a-days to stage the lymphoma because of:

- the availability and accuracy of the imaging techniques such as USG, CT and MRI;
- the accuracy of the CT-guided FNAC of the liver and lymph nodes;
- the significant morbidity (about 10%) from wound infection, subphrenic abscess, pulmonary infection and embolism, gastrointestinal haemorrhage;
- Splenectomy increases the risk of overwhelming bacterial sepsis in children below 15 years of age;
- the association between splenectomy and acute myeloid leukaemia noted in patients receiving chemotherapy;
- the increased morbidity which is upto 6%.

Staging laparotomy is also not justified in stage III & IV Hodgkin's lymphoma and in most cases of non-Hodgkin's lymphoma as they are treated by combination chemotherapy in most centres.

Clinical staging (Ann Arbor staging)**Stage I**

- Involvement of a single lymph node region e.g. palpable left supraclavicular nodes.

OR

- Involvement of a single extralymphatic organ or site (IE).

Stage II

- Involvement of two or more lymph node regions on the same side of the diaphragm – above or below e.g. left supraclavicular and left axillary nodes or left supraclavicular, left axillary and right supraclavicular nodes.

OR

- Involvement of one extralymphatic organ or site with one or more lymph node regions on the same side of the diaphragm (IIE).

Stage III

- Involvement of lymph node regions on both sides of diaphragm e.g. left supraclavicular and left inguinal nodes.

±

- Involvement of spleen (IIIS) or involvement of one extralymphatic organ or site (IIIE) or both (IIIE & S).

Stage IV

- Diffuse or disseminated involvement of extralymphatic organs or sites.

±

- Lymph node involvement.

The suffix (E) indicates an extralymphatic site or organ. The suffix (S) indicates splenic disease.

Extralymphatic site or organ involves lung, pleura, liver, bone and bone marrow, skin and subcutaneous tissue.

All stages are further subdivided into groups A or B on the basis of the absence (A) or presence (B) of the associated generalised symptoms such as weight loss, fever, night sweats, anaemia.

The clinical staging of HL is required to determine accurately the extent of the disease, the subsequent treatment and prognosis.

Differential diagnosis

From other causes of generalised or localised lymphadenopathy:

1. Tuberculosis (commonest cause of lymphadenopathy in our country).
2. Secondaries (second most common cause of lymphadenopathy in elderly people).
3. Brucellosis.
4. Toxoplasmosis.
5. Sarcoidosis.
6. Infectious mononucleosis.
7. Leukaemias.

Treatment

Two forms of treatments are available and they are stage and site dependent: *Radiotherapy* and *combination chemotherapy*.

Surgery has no role except lymph node biopsy to confirm the diagnosis, the stage and grade of the tumour. Staging laparotomy is less frequently used now-a-days because of increased morbidity.

Radiotherapy is the treatment of choice in stage I, II, and IIIA diseases.

Combination chemotherapy is used in stage IIIB and IV diseases.

Radiotherapy**Indications**

- Stage I, II & IIIA diseases.

Type of machine

- Linear accelerator is preferred than cobalt 60, because it provides sharply localised radiation.

Dose

- Total dose of 35–40 centigray (cGy) given over a period of 4 weeks (5 days each week).

Mode of delivery

Differs in various stages (Fig. 4.4):

- (a) *Stage IA with lymphocyte predominant* can be treated by Involved Field (IF) radiotherapy.
- (b) *Stage IA with other histology*
Stage IIA with any histology
Treated by Extended Field (EF) radiotherapy
- (c) *Stage IB, IIB and IIIA* – treated by a technique called Total Axial Nodal Irradiation (TANI) which includes a combination of Mantle field and Inverted Y field.

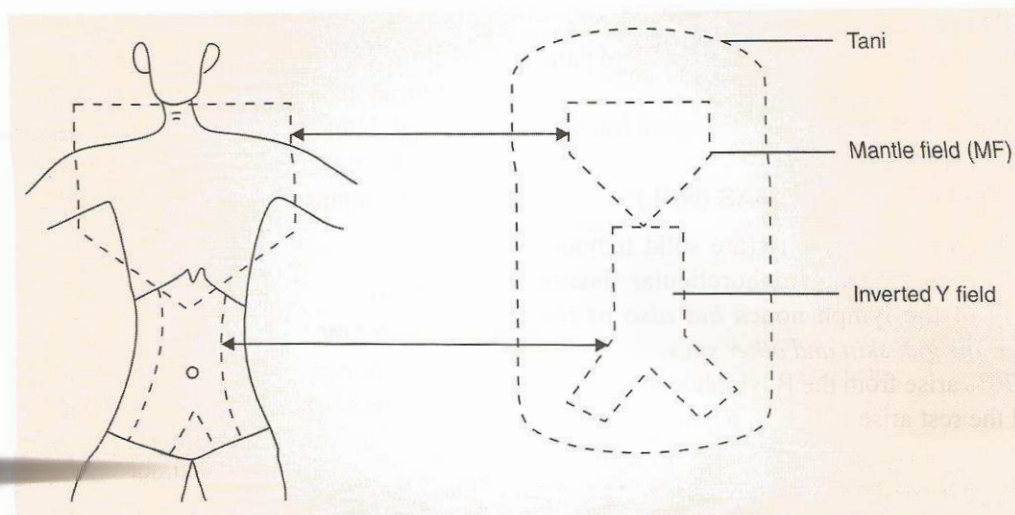


Fig. 4.4. Mode of delivery of radiotherapy in lymphoma.

Extended Field (EF) radiotherapy can be given as:

- Mantle field* which covered both cervicals, both axillary regions, the mediastinal and upper para-aortic nodes down to the level of L2 – used for supradiaphragmatic disease.
- Inverted Y field* – which covers para-aortic, both iliacs, inguinal and femoral areas – used for infradiaphragmatic disease.
- TANI* – includes irradiation of both Mantle and Inverted Y fields – first the mantle field followed by inverted Y field after an interval of 4–6 weeks.

(ii) Bone marrow suppression.

(iii) Infertility both in men and women.

2. ABVD regime:

- Includes Adriamycin, Bleomycin, Vinblastine, Dacarbazine.
- Less leukomogenic.
- Causes less infertility.
- Currently the preferred treatment.

3. Other regimens available are:

- BEACOPP, COPPABVD, Stanford V, MOPPBV.

Chemotherapy

Indications

1. Stage IIIB and IV diseases, because these are considered systemic diseases.
2. Stage IIIA has also been treated at some centres by chemotherapy with equal success.
3. Usually combination chemotherapy is used.

Regimens of chemotherapy

1. MOPP regime

- Includes Mechlorethamine, Oncovin (Vincristine), Procarbazine, Prednisolone.
- MOPP is administered for six two weeks cycles of chemotherapy with two weeks rest between each period of drugs administration.
- Basic regimen but is highly toxic.
- Side effects:
 - (i) Development of acute myeloid leukaemia.

Prognosis

Depends on:

- Extent of disease (Stages I, II, III, IV)
- Presence of symptoms (B – presence of symptoms; A – absence of symptoms)
- Histology (REAL classification)

5 years survival rates for HL

1. 85% for stage I and II
2. 70% for stage IIIA
3. 50% for stage IIIB
4. 40% for stage IV

Cause of death

Usually from

1. Systemic infection like respiratory tract infection, fungal, bacterial e.g. tuberculosis because of immunosuppression.
2. Systemic failure.

3. Bone marrow failure.
4. Acute spinal cord compression leading to paraplegia.
5. Multiple bone metastasis, pathological fracture.

NON-HODGKIN'S LYMPHOMAS (NHL)

The non-Hodgkin's lymphomas are solid tumours arising in the peripheral lymphoreticular tissues particularly of the lymph nodes *but also of the oropharynx, the gut, skin and other sites.*

65% to 70% arise from the B lymphocytes (B cell NHL) and the rest arise from the T lymphocytes (T cell NHL):

- Rarely NHL arises from histiocytes.
- *RS cells are absent in NHL.*

Rappaport classified NHL morphologically into 4 types, each of which can be nodular (follicular) or diffuse.

1. Well-differentiated lymphocytic.
 2. Poorly differentiated lymphocytic.
 3. Mixed lymphocytic and histiocytic.
 4. Histiocytic (reticulum cells).
- Prognostically these are conveniently divided into:

Morphological type	Degree of malignancy
1. Small lymphocytes and a nodular structure	Low grade tumour
2. Large lymphocytes with small lymphocytes, histiocytes and a diffuse structure	High grade tumour

- NHL has a non-continuous spread while HL has an orderly continuous spread.
- NHL has a more frequent involvement of multiple peripheral nodes compared to HL which often remains localised to one group of nodes.
- The Waldeyer's ring, epitrochlear nodes, and mesenteric nodes involvement are common in NHL, while their involvement is rare in HL.
- B symptoms described for HL are less common in patients with NHL.
- Prognosis and curability—poorer than HL.

Clinical presentation

1. Usually occurs in the 60–80 years age group.
2. Lymphadenopathy is a common presenting feature and usually generalised.

There is more chance of involvement of epitrochlear and Waldeyer's ring.

3. Extranodal issue is frequently involved such as orbit, intestine, breast, testis.
4. There may be generalised spread to involve many organs e.g. CNS, kidney, liver, lungs and heart.

Treatment

Chemotherapy

- *Chemotherapy is the mainstay of treatment.* Regimens of chemotherapy in use are:
- COP regime includes Cyclophosphamide, Oncovin (vincristine) and Prednisolone.
- CHOP regime includes Cyclophosphamide, Hydroxydaunorubicin, Oncovin and Prednisolone.
- Other regimes include BACOP (Bleomycin Adriamycin + COP).
- *Chlorambucil as a single drug oral therapy* may be used in low grade NHL.

Radiotherapy

- In some centres, *patients with low grade stages I & II NHL have also been treated with involved field (IF) radiation.*
- Following radiotherapy, the local recurrence rate is nearly nil with nodular lymphoma and around 15% with diffuse lymphoma.
- Whole body radiation (WBR) may be used in cases after failure of chemotherapy.

Monoclonal antibody therapy

- Recently being used in some centres with improved success rates.
- *It is the use of monoclonal antibodies (mAb) to specific target cells (antigens).* The main objective is stimulating the patient's immune system to attack the malignant tumour cells (antigens) and thus prevention of tumour growth by blocking specific cell receptors. Following FDA approved therapeutic antibodies are in use for non-Hodgkin's lymphoma: Tositumomab, Ibritumomab, Rituximab.

BURKITT'S LYMPHOMA

- It is a type of *high grade non-Hodgkin's lymphoma.*
- It is named after Burkitt, a surgeon who first described the disease while working in Africa.
- It is caused by Epstein-Barr virus (EBV).

Comparison between HL and NHL

	HL	NHL
Age	Bimodal age	> 60 years of age
Sites of disease		
• Nodal involvement	Common (90%) but localised to one group, commonly cervical group	Common (60%) but involves multiple groups of lymph nodes
– Mediastinal	Common	Uncommon
– Abdominal	Uncommon	Common
– Epirochlears	Uncommon	Common
– Waldeyer's	Uncommon	Common
• Nodal spread	Contiguous	Non-contiguous
• Extranodal involvement	Uncommon	Common
Cells of origin	B lymphocytes (90%)	B lymphocytes (60%) T lymphocytes Null cells Histiocytes
Systemic symptoms	Common	Uncommon
• Fluctuating fever	Present	Absent
• Alcohol-induced pain or pruritus	Characteristic	Absent
Treatment	Radiotherapy for stages I & II diseases Combination chemotherapy for stages III & IV diseases	Poorer than HL Combination chemotherapy mainly for all stages
Prognosis	Better than NHL	Poorer than HL

- It is believed that *impaired immunity* provides an opening for development of EB virus infection. (EBV also causes glandular fever – infectious mononucleosis, a non-malignant condition in young adults in Western societies).
- Clinically Burkitt's lymphoma can be divided into 3 clinical varieties: endemic, sporadic and immunodeficiency associated variants.

(a) Endemic variety

- Occurs in equatorial Africa, Nigeria, New Guinea and South America, *where malaria is endemic*.

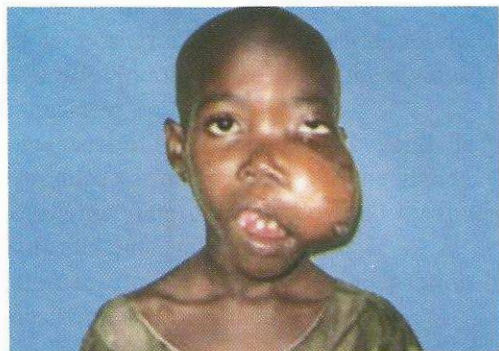


Fig. 4.5. Burkitt's lymphoma involving left maxilla.

Where malaria has been eradicated the incidence of Burkitt's lymphoma falls suggesting a possible insect vector of an infective origin.

- *Males* are predominantly affected.
- *Young children are affected more*—over 80% of cases occur between the ages of 3 and 12 years.
- The tumour characteristically involves the jaw or the facial bones (about 60%). Maxillary lesions may present as oral or orbital tumours.
- The tumour grows rapidly and is soft and painless.
- The disease may involve the distal ileum, caecum, ovaries, kidneys and breasts.
- *Endemic variety has a good prognosis.*

(b) Sporadic variety

- Occurs throughout the world.
- Jaw is less commonly involved compared to the endemic variety. Ileo-caecal region is the common site of involvement.

(c) Immunodeficiency variety

Usually associated with HIV infection. Burkitt's lymphoma can be an initial manifestation of AIDS.

Histology: Histological appearance is typical and striking—presence of dense masses of lymphocytes interspersed with large histiocytes or macrophages containing debris derived from dead body of apoptotic tumour cells, contributing the “starry-sky” appearance.

Treatment

- *Endemic variety* responds well to chemotherapy.
 - (i) Single drug cyclophosphamide has been shown to be effective.
 - (ii) Long-term survival without maintenance therapy has been reported.
 - (iii) A number of spontaneous remissions have also been recorded.
- *Sporadic variety* needs to be treated with combination chemotherapy.
- Other treatments like immunotherapy, bone marrow transplants, surgery to remove the tumour, and radiotherapy may also be required.

5

CHAPTER



Salivary Glands

SURGICAL ANATOMY

There are three pairs of major salivary glands – the parotid, the submandibular and the sublingual glands. Besides these main salivary glands, small accessory glands are found scattered over the palate, lips, cheek, tonsil and tongue. These glands are occasional sites for development of mixed salivary tumour.

Parotid gland (Fig. 5.1)

It is the largest of the three paired salivary glands. It is situated in the parotid mould. The mould presents the following boundaries:

- *Anteriorly:* Posterior border of the ramus of the mandible and the muscles attached to it – superficially, the masseter, and on the deeper aspect, the medial pterygoid muscle.
- *Posteriorly:* Mastoid process and sternomastoid muscle.
- *Above:* By the zygomatic arch.
- *Below:* Posterior belly of digastric muscle.
- *Medially:* Styloid process and the styloid groups of muscles.

Some parts of the gland extend beyond the mould and produce various processes of the gland.

Coverings: The gland is covered by inner true capsule and outer false capsule.

True capsule is formed by condensation of the fibrous stroma of the gland.

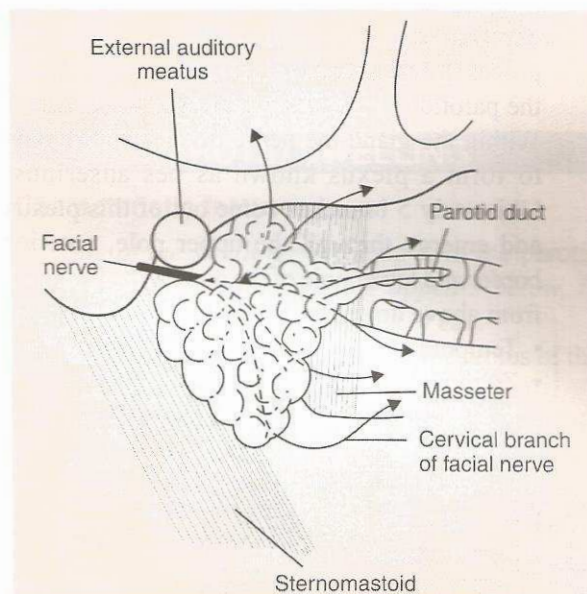


Fig. 5.1. Parotid mould with the gland. Note that the parotid duct opens into the cheek opposite the crown of the upper second molar tooth.

False capsule is known as *parotid sheath* and is formed by the splitting of the investing layer of deep cervical fascia. The superficial lamella of the sheath is strong and attached above to the zygomatic arch; and anteriorly it extends forward over the masseter muscle to form the parotidomasseteric fascia which is dense and tough.

The deep lamella is thin. It passes deep to the gland and is attached to the tympanic plate and styloid process of the temporal bone. Anteroinferiorly this deep lamella is thickened and known as stylo-mandibular ligament. It separates the parotid from the submandibular gland.

Note: It must be remembered that the parotid fascia is so thick and tough that the parotid abscess does not show fluctuation until very late.

Structures passing through the parotid gland

The structures are arranged in three zones:

Superficial zone – Occupied by the nerves

1. Facial nerve: It emerges from the stylomastoid foramen, and winds laterally to the base of the styloid process (so, can be exposed surgically in the inverted V between the bony part of the external auditory meatus and the mastoid process). Just beyond this point the nerve enters the parotid gland at its posteromedial surface. Within the gland the nerve divides and rejoins to form a plexus known as pes anserinus. Ultimately 5 branches come out of this plexus and emerge through the upper pole, anterior border and lower pole of the gland. The branches from above downwards are (Figs. 5.1 and 5.3):
 - Temporal
 - Zygomatic
 - Buccal
 - Mandibular
 - Cervical
2. Great auricular nerve.
3. Auriculotemporal nerve.

Intermediate zone – Occupied by the veins

1. Retromandibular vein which is formed by the union of superficial temporal and maxillary veins.
2. Anterior division of retromandibular vein which joins the anterior facial vein to form the common facial vein.
3. Posterior division of retromandibular vein which joins the posterior auricular vein to form the external jugular vein.

Deep zone – Occupied by the arteries

1. External carotid artery dividing into superficial temporal and maxillary arteries.

2. Transverse facial artery – a branch of superficial temporal artery.
3. Sometimes posterior auricular artery arises within the gland, from the external carotid artery.

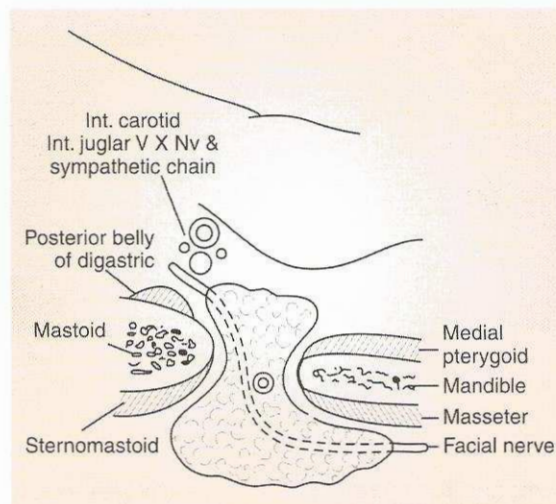


Fig. 5.2. Zones of parotid gland and its surrounding structures. Note that the facial nerve is the most superficial of the structures traversing the gland.

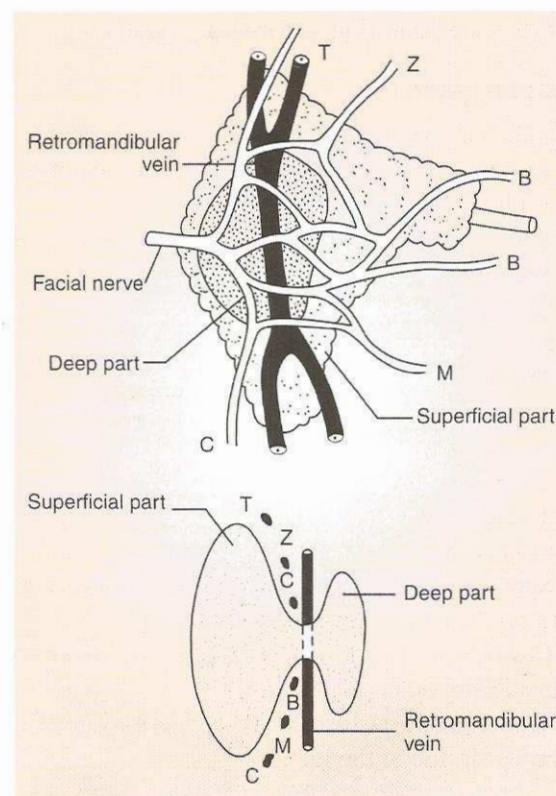


Fig. 5.3. Patey's faciovenous plane.

Patey's faciovenous plane (Figs. 5.2 and 5.3)

Within the gland the facial nerve and its branches lie with remarkable constancy in one plane *superficial to the retromandibular vein*. The plane in which the nerve and the vein lie has been designated by Patey as the faciovenous plane. In this plane the gland can be split sagittally into two parts – superficial and deep. *Along this plane superficial parotidectomy is carried out without injuring the nerve.*

The parotid duct emerges from the anterior border of the gland and runs horizontally across the masseter a finger breadth below the zygomatic arch and it makes a right-angled turn at the anterior border of masseter to pierce the buccinator to open in the vestibule of the mouth (buccal aspect of the cheek) via a small papilla opposite to the crown of the upper second molar tooth.

Nerve supply of parotid gland

Secretomotor supply via the auriculotemporal nerve carrying the parasympathetic fibres from the inferior salivary nucleus via the otic ganglion.

Submandibular gland

The gland is situated in the submandibular region partly below the mandible and partly deep to it. Each gland consists of a larger superficial part and a smaller deep part: both the parts are continuous around the posterior border of mylohyoid muscle. The gland is enclosed in a loose sheath derived from the investing layer of deep cervical fascia. Superficial part of the gland lies on the mylohyoid, hyoglossus and the middle constrictor muscle from before backwards. The deep part extends in the interval between mylohyoid and hyoglossus up to the sublingual salivary gland.

Relations of the deep part

- Laterally – Mylohyoid.
- Medially – Hyoglossus.
- Below – Hypoglossal nerve accompanied by a pair of veins.
- Above – Lingual nerve and the submandibular ganglia. The lingual nerve hooks the submandibular duct from superficial to deep at its lower border.

Superficially the gland is covered by the platysma and it is crossed by the cervical branch of the facial nerve and by the anterior facial vein.

The facial artery comes in close relationship with the gland. It approaches the gland posteriorly and then arches over the superior aspect (which it indents) to attain the inferior border of the mandible and thence to ascend onto the face in front of the masseter.

The submandibular duct (Wharton's duct) emerges from the medial surface of the gland and runs forward under the cover of mylohyoid beneath the mucosa of the floor of the mouth along the side of the tongue to open on the sublingual papilla at the side of the frenum. Here its orifice is readily visible and saliva can be seen trickling from it. (A stone in the Wharton's duct can be felt bimanually in the floor of the mouth and often can be seen if sufficiently large.)

The sublingual gland lies immediately lateral to the duct.

The submandibular lymph nodes lie superficial to the submandibular gland. They may lie partly embedded within the gland.

PAROTID SWELLING

Characteristic features

1. *Site of the swelling:* Swelling is in the parotid region or mould. *Swelling appears below, in front of and behind the lobule of ear.*
2. It obliterates the furrow behind the ramus of the mandible.
3. Shape of the swelling may look like parotid gland if it is a larger one.
4. In many cases where the swelling is big, the lobule of the ear is pushed up.

Clinical examination of parotid swelling

In addition, to note the site, size, shape, consistency, etc., of the swelling, the following points must be examined:

1. Relation of the swelling to the skin – Free or fixed.
2. Relation of the swelling with relaxed and taut masseter (*the patient is asked to clench the teeth thus making the masseter taut*) – Mobile or fixed.
3. Examination of facial nerve function:
To rule out facial nerve palsy following will be noted:
 - Observe the nasolabial fold and furrows of the brow – less marked on the side of facial nerve palsy.

- **Test for motor function:** Ask the patient
 - *To show his clenched teeth:* When the angle of mouth will be drawn to the normal side.
 - *To puff out the cheeks:* The paralysed side bellows out more than the normal side.
 - *To shut his eyes:* The patient will not be able to close the eye on the affected side. Moreover the eyeball will be seen to roll upwards.
 - *To move his eyebrows upwards to wrinkle the forehead:* The paralysed side remains immobile.
- 4. **Intraoral examination:**
 - Approximately 10% of parotid tumours develop in the deep lobe and lie beneath the facial nerve.
 - So, examine the oral cavity. The deep lobe tumour presents with a parapharyngeal mass which pushes the pharyngeal wall, tonsil and soft palate medially.
 - Deep lobe tumours may present with the complaints of dysphagia.
 - Deep lobe tumours usually do not appear as a gross swelling on the surface of the face until it is too large or both the superficial and deep lobes are involved.
 - Parotid duct and its orifice – for redness, oedema of the duct, or exudation of pus.
- 5. Movements of the temporomandibular joint.
- 6. Lymph nodes of the neck.

Differential diagnosis of the swelling in the neighbourhood of parotid region

I. Pre-auricular lymphadenitis

Diagnostic points:

1. Ovoid, firm, smooth, swelling is situated just in front of the tragus.
2. The furrow behind the ramus of the mandible is not obliterated.
3. Presence of some primary focus of infection or tumour in the drainage area – forehead and temporal region of the scalp, auditory meatus, eyelids, cheek.
4. The swelling is very mobile, *as the node is outside the capsule of the parotid gland* – the most distinctive physical sign. (Most tumours of the parotid gland are tethered to the gland. So, they are less mobile.)

II. Swelling in connection with masseter muscle

1. Fibroma
2. Lipoma
3. Rhabdomyoma
4. Idiopathic hypertrophy of the masseter muscle.

Diagnostic points: The swelling becomes fixed and hard beneath the fingers when the masseter is contracted (by clenching the teeth) and free and relatively soft when the masseter is relaxed.

III. Swelling in relation to the overlying skin, e.g., sebaceous cyst, lipoma and haemangioma.

IV. Neuroma of the facial nerve.

V. Adamantinoma of the mandible.

NEOPLASMS OF THE SALIVARY GLANDS

Approximately 75% of the neoplasms of the salivary gland occur in the parotid glands. 80% of parotid tumours are benign and of these, pleomorphic adenoma is the commonest (80%) and Warthin's tumour is the second most common benign tumour. The remaining 20% are malignant.

About 15% of salivary gland tumours occur in the submandibular salivary glands, 60% of which are benign. Of the benign tumours 90% are pleomorphic adenomas.

10% of the salivary tumours occur in the sublingual salivary glands and minor salivary glands situated in the palate, lips and cheeks. Of these 40% are benign and virtually all these benign tumours are pleomorphic adenomas.

PAROTID TUMOUR

The International classification

- I. Epithelial tumours.
- II. Non-epithelial tumours.
- III. Metastatic carcinoma.

I. Epithelial tumours

A. Adenomas

- (a) Pleomorphic adenoma (myoepithelial origin) also known as mixed parotid tumour – potentially malignant.

Note: Pleomorphic means many forms.

- (b) Monomorphic adenomas (duct cell origin):
 1. Adenolymphoma or Warthin's tumour
 2. Oxyphilic adenoma
 3. Other types

- B. Mucoepidermoid tumour (arising from duct cells) – Low grade malignancy and very slowly growing tumour.
- C. Acinic cell tumour (arising from acinar cells)
- D. Carcinoma:
 - (a) Adenoid cystic carcinoma or cylindroma (arising from glandular cells) – relatively radiosensitive.
 - (b) Adenocarcinoma (arising from glandular cells).
 - (c) Epidermoid or squamous cell carcinoma (arising from duct cells).
 - (d) Undifferentiated carcinoma:
 - Carcinoma in pleomorphic adenoma.
 - Anaplastic carcinoma.

II. Non-epithelial tumours

- (a) Haemangioma
- (b) Lymphangioma
- (c) Neurofibroma.

III. Metastatic carcinoma

- Secondary to the epidermoid carcinoma and malignant melanoma.

Characteristic features

1. Most benign adenomas (90%) are superficial to the facial nerve. The rest are deep lobe tumours.
2. Like benign tumours, malignant tumours also usually present as painless, slowly growing lump.
3. Presence of pain, facial nerve involvement or fixation of the lump to adjacent structures indicate a more advanced stage with a worse prognosis.
4. *No formal incisional biopsy should be performed in parotid tumour, except in tumour of ectopic salivary glands, e.g., palatal salivary gland.*
5. *Recently FNAC using 23 G needle is advocated as it does not cause tumour cells implantation in the needle tract..*
6. For deep lobe tumour, FNAB through intraoral route is recommended and it does not cause surface skin infiltration.
7. CT or MRI are used if the lesion is large or involves deep lobe or adjacent structures.
8. All the parotid neoplasms are *radio-resistant*. So, complete excision with negative margins is the treatment of choice.

ADENOLYMPHOMA

(Syn.: Papillary cystadenoma lymphomatosum; Warthin's tumour)

It is an epithelial tumour arising within a **periparotid** lymph node containing ectopic salivary tissue.

During development of the parotid gland, the ductal structures from the primary parotid bud (*containing epithelial cells*) may grow into the developing juxta-parotid lymph nodes. These epithelial cells later may proliferate in an acinar form and give rise to a swelling called adenolymphoma.

It is usually limited to the area of the parotid gland because lymph node containing ectopic salivary tissue is usually found in the parotid gland but not in other submaxillary salivary glands.

Pathology

1. This is a cystic tumour containing epithelial and lymphoid tissues.
2. The neoplasm is situated outside the parotid capsule, or just beneath the capsule embedded in the superficial lobe.
3. Benign, slowly growing, encapsulated tumour. Does not undergo malignant change.
4. May be multiple or bilateral.
5. Though developmental in origin, it does not appear before the age of forty years.

Macroscopy: Multiple cysts are revealed varying in size from pin-head to hazel nut. The cyst usually contains glairy brown mucus.

Microscopy: Papillary epithelium embedded in a lymphoid stroma – an extremely characteristic feature.

Diagnostic criteria

1. **Age:** Commonly in elderly people after 40 years.
2. **Sex:** Males are more commonly affected.
3. **Race:** Mostly confined to white races. It has never been recorded in Negros.
4. The patient presents with *a slowly growing painless swelling on the side of the face*.
5. **Site:** The swelling is situated in the parotid region commonly at or below the level of the lower border of the mandible.
6. **Shape and size:** Oval or circular, and **usually** 1–3 cm in diameter.



Fig. 5.4. Parotid swelling left side. Smooth, soft swelling below and behind the lobule of the ear (typical site)—adenolymphoma.



Fig. 5.5. Adenolymphoma. A smooth, soft swelling in the parotid region over the angle of the mandible. No fixity to the overlying or underlying structures. D/D: 1. Mixed parotid tumour—can be differentiated by ^{99m}Tc pertechnetate scan. *Adenolymphoma produces a 'hot spot'.* 2. Parotid cyst—aspiration will confirm fluid.

7. Surface: Smooth with well-defined margins.
8. Consistence: *Soft to cystic*, so often fluctuating. *Not translucent.*
9. Not tender.
10. Overlying skin is free from the swelling and looks and feels normal. There is no evidence of venous prominence.
11. *Mobility*: Not fixed to the deeper structures, i.e., masseter, so mobile in both axes.
12. *No evidence of involvement of the facial nerve.*
13. Cervical lymph nodes are not enlarged.

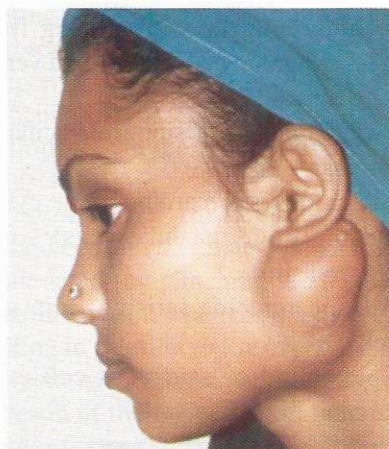


Fig. 5.6. Left parotid tumour. Smooth, soft swelling below and behind the lobule of the ear over the angle of the mandible (a typical location); the ear lobule is elevated—adenolymphoma. No evidence of facial palsy noted. (Courtesy: Prof. Sudeb Saha, MS, FAMS)

Differential diagnosis

1. Sometimes adenolymphoma may be confused with mixed parotid tumour because of presentation at the same age group. *So they can be differentiated by ^{99m}Tc pertechnetate scan—the adenolymphoma produces a 'hot spot' while all other neoplasms form 'cold spot', so that a firm pre-operative diagnosis is possible without biopsy.*

However, the treatment of both the conditions is superficial parotidectomy.

2. Sometimes the swelling may be situated in the neck below the angle of the mandible when it is to be differentiated from:
 - (a) Branchial cyst (aspirated material will show cholesterol crystals).
 - (b) Enlarged cervical lymph node.
3. Parotid cyst.

Treatment

Superficial parotidectomy.

PLEOMORPHIC ADENOMA (Syn.: Mixed parotid tumour)

It is the most common tumour of the parotid gland. It is called mixed tumour because it contains

pleomorphic stroma e.g. cartilaginous tissue beside epithelial tissue. *Pleomorphic means many forms.*

It is a benign tumour but may change into malignancy at any time and hence should be considered as potentially malignant.

Pathology

1. Origin – Regarding its origin there are two views:
 - (i) According to some, the mixed parotid tumours arise from the embryonic rests due to invagination of the oral ectoderm along with the salivary and oral glands.
 - (ii) According to others, the tumours are true adenomas of the salivary glands rather than true mixed tumours.
2. Slowly growing benign tumour, gradually increasing in size over a period of many years.
3. *Unicentric in origin. But recurrence is multicentric.*
4. The tumour possesses a pseudocapsule. The tumour tissue juts out if incision is made on the capsule.
5. *Potentially malignant*, therefore, malignancy can supervene at any time.
6. Some fringe of the tumour mass may protrude through the thinned out capsule and invade the adjoining areas. These protrusions through the capsular deficiencies may give rise to recurrence after removal of the tumour.
7. *The tumour is radioresistant.*

Histological picture

It is adenoma of the salivary gland with a *pleomorphic stroma* containing fibrous, lymphoid, myxomatous, pseudocartilaginous elements in addition to epithelial elements. It is believed that the cartilage it contains is not mesodermal in origin but is mucin or pseudomucin secreted by the epithelial cells which when stained resemble cartilage.

Diagnostic criteria

1. *Age*: Usually first appears in adult life.
2. *Sex*: More common in males.
3. The patient presents with a *slowly growing painless swelling on a side of the face*, which has been existent for months or years without any appreciable change in size.

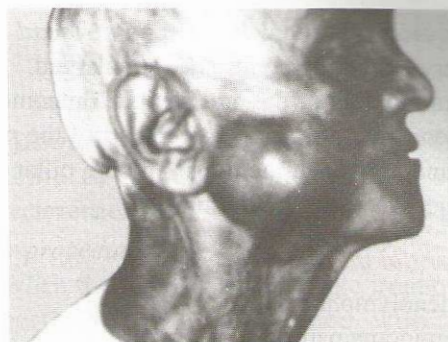


Fig. 5.7. Mixed parotid tumour right side (typical location). No facial nerve involvement.



Fig. 5.8. Swelling over the left parotid region extending below and behind the ear lobule (typical location). No facial nerve involvement – mixed parotid tumour.

4. *Site*: The swelling is situated in the parotid region, usually commences in the part of the parotid gland overlying the angle of the mandible.
5. *Shape*: Rounded swelling with well-defined margins.
6. *Size*: Varies from a pea-sized nodule to a large, almost pendulous mass.
7. *Surface* is smooth, sometimes lobulated.
8. *Consistence*: *Firm elastic to rubbery hard, not fluctuant, not translucent.*
9. Not tender.
10. Overlying skin is free from the swelling, and looks and feels normal. There is no venous prominence.
11. *Mobility*: Not fixed to the deeper structure, e.g., masseter. It is mobile in both axes over both the relaxed and taut masseter.

12. No evidence of involvement of the facial nerve.
13. Cervical lymph nodes are not enlarged.
14. Examination of the oral cavity: A pleomorphic adenoma arising in the deep part of the parotid gland may push the tonsil and the pillar of the fauces towards the midline.

Differential diagnosis

1. Adenolymphoma.
2. Carcinoma parotid.

Complications

1. Malignant changes:
 - Occasionally the variegated epithelial elements may burst into mitotic activity giving rise to pleomorphic carcinoma.
 - On rare occasions, one epithelial component alone runs a riot resulting in highly malignant anaplastic carcinoma.
2. Recurrence: Particularly after enucleation. *Recurrence is multicentric and may be malignant.*

Causes of recurrence may be:

 - Incomplete surgical excision.
 - Implantation of the tumour cells during manipulation.
 - Multicentric growth potentiality of the tumour.

Treatment

The tumour is *radioresistant*.

So, *treatment of choice is surgery*. The surgical procedure includes *superficial parotidectomy with preservation of the facial nerve*, because most of the tumours lie in the superficial lobe. The operation is known as *Patey's superficial parotidectomy*.

The dissection is made in the Patey's faciovenous plane, and commenced at the posteroinferior border of the parotid gland, where the main trunk of the facial nerve is found out.

Incidence of transient facial nerve palsy due to neuropraxia may occur in upto 30% cases but usually recovers within 6 weeks.

Criteria of malignant change in a mixed parotid tumour

1. Sudden and rapid increase in the size of a swelling which is present for long times.
2. Painless tumour becomes painful and so tender.

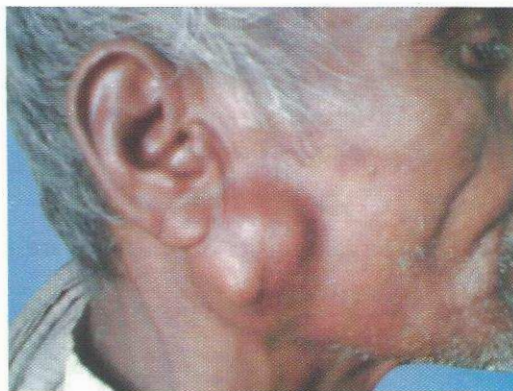


Fig. 5.9. Right parotid tumour with multiple nodules over the angle of the mandible (a typical location); no facial nerve involvement – mixed parotid tumour. (Courtesy: Prof. Sudeb Saha, MS, FAMS)

3. Feels stony hard. Surface may be nodular.
4. The growth becomes fixed to the deeper structures, e.g., masseter, mandible.
5. Overlying skin may become fixed to the swelling and looks and feels reddish-blue and hot. There may be fungation and ulceration of the skin.
6. Evidence of facial nerve involvement causing asymmetry of the face and difficulty in closing the eyes.
7. Areas of anaesthesia over the skin.
8. Jaw movements may become restricted.
9. Veins over the swelling become prominent.
10. Enlargement of cervical lymph nodes may be present.
11. There may be evidence of disseminated blood-borne metastases.

Note: There is no place of incisional biopsy in parotid tumour as chances of seedling and recurrence are high and also there is chance of injury to the facial nerve while doing the biopsy. Recently fine needle aspiration cytology (FNAC) using 23G needle has been advocated by few surgeons. For deep lobe tumours FNAB through intraoral route is advocated.

CARCINOMA OF THE PAROTID GLAND

Characteristic features

1. Malignancy may start *de novo* in the parotid gland with a rapidly increasing swelling from the onset.

2. Malignant change may occur in a mixed parotid tumour which has pursued an apparently benign course for many years with history of recent rapid increase of a pre-existing benign swelling.
3. Usually arises in elderly persons.
4. Men and women are equally affected.
5. Evidence of facial nerve palsy is most likely. So perform examination for the facial nerve palsy.
6. The lymph nodes are involved in 15 percent cases.
7. The tumour is radioresistant.

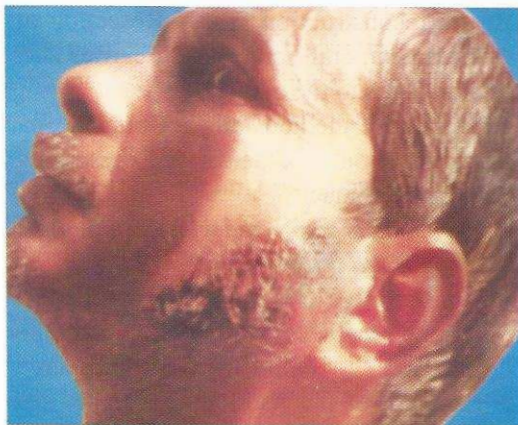


Fig. 5.10. Carcinoma of the parotid gland infiltrating the overlying skin – the facial nerve was not involved.

Diagnostic criteria (Fig. 5.10)

1. Vide before (malignant change in a mixed parotid tumour - see page 124).
2. FNAC from the swelling to confirm diagnosis.
3. CT scan or MRI of parotid region to assess the tumour extent.

Treatment

Malignant parotid tumour is radioresistant. So, treatment of choice is surgery. The procedures include:

- A. **Complete parotidectomy** which may be either:
 1. *Radical parotidectomy* (i.e., where the facial nerve is sacrificed, particularly if there is a reasonable prospect of cure. For, it is otherwise certain that nerve function will eventually be destroyed by the tumour).
 2. *Total conservative parotidectomy* i.e., total parotidectomy with preservation of facial nerve. Superficial parotidectomy is done first

preserving the facial nerve; the deep part of the gland is then removed preserving the facial nerve branches.

Postoperative radiotherapy is advocated by some to minimise the chances of local recurrence.

Other procedures:

- B. When the tumour is as a whole irremovable—partial excision combined with radium implantation may give the best prospect of palliation.
- C. Block dissection of the lymph nodes usually on the side of the tumour is occasionally of value when operable metastatic lymph nodes are present after successful treatment of the primary tumour.
- D. If the tumour is fixed to the deeper structures and found to be inoperable — only palliative radiotherapy is advocated.
- E. Local recurrences should be treated by radiotherapy.
- F. Role of chemotherapy: *Chemotherapy has limited role.* Methotrexate or 5-fluorouracil therapy may cause limited regression, but its efficacy is not established for long-term palliation.

Perfusion with cyclophosphamide through the superficial temporal artery has produced marked regression in certain cases of advanced parotid carcinoma.

PAROTID FISTULA

A salivary gland fistula may be *internal* or *external*. An internal fistula which opens inside the mouth does not give rise to symptoms, and hence is not of much clinical importance.

An external fistula of the submandibular salivary gland is very rare and, if it occurs, can be cured readily by removal of the gland. But an external fistula of parotid causes troublesome condition, and hence is of much importance.

Parotid fistula may be *gland fistula* or *duct fistula*.

Causes

1. Rupture of parotid abscess.
2. Inadvertent incision during drainage of parotid abscess.

3. Penetrating injury, especially by glass splinters.
4. Aftermath of operations on the gland, e.g., superficial parotidectomy — a persistent fistula is rare, though some leakage of saliva occurs for several days.

Diagnostic criteria

1. *When the external fistula is connected with the gland:*
 - There will be little moisture on the face at the time of eating.
 - The external opening is pinpoint.
 - It produces little inconvenience.
 - Though there is discharge for several months, it usually closes spontaneously.
2. *When the external fistula is connected with the duct, particularly a major duct:*
 - There is outpouring of parotid secretions onto the cheek every time during eating, smelling of food, thought of taking food.
 - Excoriation of the neighbourhood skin.
 - So, there will be extreme discomfort.

Investigation

A sialogram with a watery solution of lipiodol must be done in these cases to assess whether the fistula is in relation to the main duct or a ductule. This information will indicate the proper line of treatment.

Treatment

- A. *If the fistula is connected with the main duct* — Reconstruction of the duct after the method of Newman & Seabrook's operation.

Steps of reconstruction:

1. One probe is introduced through the orifice of the parotid duct in the vestibule of the mouth.
2. Another probe is introduced through the external fistula.
3. With the probes in position, a suitable horizontal incision is made over the cheek.
4. The ducts are dissected out and the severed ends of the duct are identified and made free.
5. A twisted tantalum wire (No. 10 size) is negotiated in both the distal and proximal parts of the duct.
6. Any portion of the twisted wire that lies

exposed is buried with the adjacent connective tissue by catgut sutures.

7. The skin is closed.
8. The distal end of the wire is suitably bent around the angle of the mouth and anchored there by adhesive plaster.

Post-operative management:

- Antibiotic to be given for several days.
 - Maintenance of oral hygiene.
 - Each day the wire splint is moved very gently.
 - The wire is to be kept in situ for about three to four weeks.
- B. If the reconstruction fails — Resection of the auriculotemporal nerve which carries secreto-motor fibres to the gland.
 - C. If the above method fails — Complete parotidectomy should be performed preserving the facial nerve intact.
 - D. *If the fistula is connected with the minor branch of the parotid gland:*
 - Conservative treatment with propantheline bromide (parasympatholytic) 50 mg 6 hourly for a week to diminish the parotid secretion.
 - Excision of the fistulous tract may also be tried.

AURICULOTEMPORAL SYNDROME

(Syn.: Frey's syndrome)

This syndrome usually follows the injury to the auriculotemporal nerve due to inadvertent incision during operation for parotidectomy, parotid abscess or suppurative parotitis.

This syndrome is characterised by the following:

- Painful sweating on the cheek of the operated side while eating (gustatory sweating).
- Sometimes cutaneous hyperaesthesia in front of and above the ear particularly during shaving.

Explanation of the syndrome

Explanations are not very satisfactory. A satisfying hypothesis is that during regeneration of the nerve after its division, there is regrowth of parasympathetic motor nerve fibres of the auriculotemporal nerve into the cutaneous nerve fibres of the skin flap. Such crossed innervation of the sweat glands produces uncomfortable gustatory sweating and hyperaesthesia.

Management

Prevention

- Proper placement of incision during parotidectomy.
- Placement of –
 - artificial membrane
 - sternomastoid muscle flap
 - temporalis fascial flap
 between the skin and the parotid bed following parotidectomy
- Resection of a segment of auriculotemporal nerve during parotidectomy.

Treatment

- Reassurance: Usually the symptoms pass off spontaneously in few months.
- Local application: Antiperspirant – Aluminium chloride is a useful astringent.
- Injection of botulinum toxin into the affected skin may help.
- Avulsion of the auriculotemporal nerve will result in effective treatment.
- Denervation by tympanic neurectomy.

Auriculotemporal nerve ascends in front of the auricle just behind the superficial temporal artery.

DISEASES OF SUBMANDIBULAR SALIVARY GLAND

CALCULUS DISEASE OF THE GLAND (SIALOLITHIASIS)

Site

Common sites are:

- Within the submandibular salivary gland substance.
- Within its duct (Wharton's duct) deep to the mucous membrane of the floor of the mouth – more common.

Submandibular salivary calculi are eighty times more common than the parotid calculi, because:

1. Submandibular duct (Wharton's duct) has a long curved and upward course and is hooked by the lingual nerve. The secretion of the submandibular salivary gland starts down in the neck and drains up in the floor of the mouth so non-dependent drainage. So there is inadequate drainage and therefore stasis which precipitates salt deposition.

Parotid duct (Stensen's duct) has a straight course and has dependent drainage.

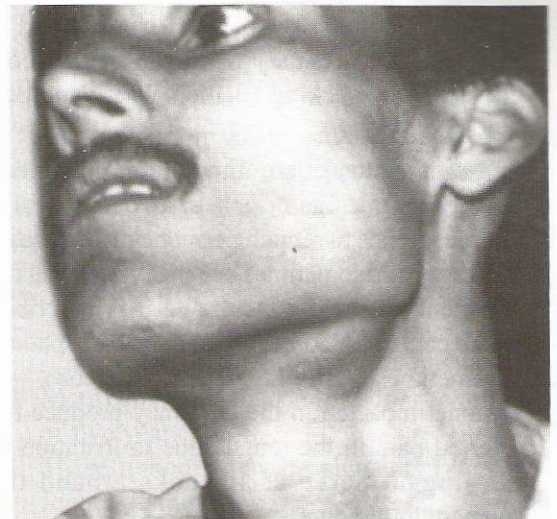


Fig. 5.11. Enlargement of the submandibular salivary gland. On bidigital palpation, there was stone in the gland itself but no stone in Wharton's duct – a case of sialolithiasis.

2. The secretion of submandibular salivary gland is more viscid and rich in salt content than parotid gland secretion.

Composition of stone

The calculus is composed mainly of the phosphate and carbonate of calcium and magnesium, with a small percentage of organic matter. It closely resembles that of tartar that collects upon the teeth. *Due to deposition of calcium salts the stone is radio-opaque.*

Cause of stone formation

Chronic inflammation of the gland – retention of secretions which later become inspissated – deposition of inorganic calcium salts on a nucleus of degenerated epithelium and inspissated secretions.

Size and shape of the stone

- The size varies from a millet to a pea.
- The shape is oval or elongated.

SUBMANDIBULAR CALCULI

Diagnostic criteria

1. Age: Mostly common in young and middle-aged adults.
2. Sex: Both sexes are equally affected.
3. The patient complains of pain and swelling at

the submandibular region (i.e., beneath and in front of the angle of the jaw), aggravated by food, classically by sucking a lemon. The gland gets tense and tender.

At first the attacks are transient and the gland quickly resumes its normal size, but in course of time it tends to remain permanently enlarged. Salivary colic sometimes occurs, typically at the commencement of a meal, and it is described by the patient as toothache (for which the patient is sometimes referred to a dental surgeon).

A stone impacted in the duct may produce the referred pain in the tongue due to irritation of the lingual nerve as it hooks around the submandibular duct.

4. **Site:** The lump is situated beneath the horizontal ramus of the mandible, 1 to 2 cm in front of the anterior border of sternomastoid muscle.
5. **Shape:** A flattened ovoid or almond-shaped.
6. **Surface:** Usually smooth but may be bosselated. The overlying skin is usually normal and freely mobile over the swollen gland.
7. **Margin:** Anterior, posterior and inferior margins of the gland are easily defined but the upper margin is impalpable as it is wedged between the mandible and the mylohyoid muscle.
8. **Consistence:** A distended submandibular gland feels firm to rubbery hard.
9. **Mobility:** Can be moved a little from side to side. The gland becomes less mobile when the muscles of the floor of the mouth are taut by asking the patient to push his tongue against the palate.
10. **Bidigital palpation:** One index finger is inserted inside the mouth between the side of the tongue and the inner surface of the alveolus. The finger of the other hand is placed under the jaw. Now first the gland and next the duct are palpated from behind forward.

By this method one can appreciate that the lump is outside the structures forming the floor of the mouth. One can palpate a stone in the deeper part of the gland or in the duct. Bidigital palpation also helps to differentiate an enlarged submandibular gland from enlarged submandibular lymph node because the finger inside the mouth can feel the deep part of the submandibular

salivary gland as it lies above the mylohyoid muscle. But, the inner finger cannot palpate the lymph node as it lies below the mylohyoid muscle.

11. **Examination of the floor of the mouth:** The patient should be asked to show the undersurface of the tongue – this will display the orifices of submandibular duct on their small papillae on either side of the frenum of the tongue. *Swollen, oedematous and congested orifice indicates the impaction of the stone. Occasionally, the stone impacted in the ampulla could be observed.*

If some lemon juice is poured in the tongue and mouth, there will be outpouring of saliva from one orifice while there will be absence of saliva flowing from the other orifice, which indicates that this duct is obstructed. At the same time one may notice the prominence or increase of the swelling in the submandibular region.

Differential diagnosis

- Submandibular lymphadenitis.
- Salivary gland neoplasm.

Investigation

All cases should be X-rayed before operation.

Submandibular salivary calculi either in the duct or in the gland substance will cast a clear radio-opaque shadow due to presence of calcium. Sometimes the stone may be non-opaque due to poor mineral content.

Treatment

Surgical

1. When the stone is in the duct: Removal of the stone is done intraorally.
 - General or local anaesthesia.
 - Tissues immediately behind the stone are grasped by the forceps which steady the stone and, thus, prevent it from slipping backwards in the gland substance.
 - An incision is made on the mucous membrane and duct directly over the stone in the long axis of the duct.
 - The stone is removed.
 - The cut ends of the mucous membrane and the duct wall are left unsutured.

2. When the stone is in the gland substance:
Excision of submandibular salivary gland.

Care to be taken during excision of submandibular gland:

- A skin crease incision is made directly over the gland.
- The posterior part of the incision should lie at least 2 cm below and in front of the angle of the mandible—to avoid injury to the cervical branch of the facial nerve.
- The facial vein and facial artery should be divided in between the ligatures twice.
- The lingual nerve and the hypoglossal nerve lying between the deep surface of the gland and mylohyoid muscle, and hyoglossus muscle should be identified and preserved. Mylohyoid muscle is retracted to remove the deep portion of the gland.

TUMOURS OF THE SUBMANDIBULAR GLAND

All the tumours involving the parotid gland may be found in the submandibular salivary gland but they are comparatively rare.

If diagnosed early, the treatment is satisfactory, because the excision of the submandibular salivary gland in toto is rather easy.

ECTOPIC SALIVARY TUMOURS

The tumours of the salivary gland may involve the tiny, unnamed minor salivary glands situated in the buccopharyngeal cavity or other uncommon sites.

Minor, unnamed salivary glands are present in the following sites:

- Hard palate
- Lips and cheeks
- Tongue
- Fauces
- Pharynx
- Larynx

Migrant (ectopic) salivary gland may be present in the following sites:

- Eyelids
- Lacrimal gland
- Ear — middle ear
- Paranasal sinuses
- Nose

- Jaws
- Skin of the face and neck.

The commonest site is the hard palate.

These tumours make up some 10 percent of all salivary gland tumours, 40 percent of which are benign and virtually all these benign tumours are pleomorphic adenoma.

Characteristic features

1. The great majority of these tumours is quite symptomless.
2. More commonly found in children below the age of five years.
3. These tumours may be pigmented (cf. Peutz-Jeghers syndrome).
4. Unless the tumour is excised early and widely, local recurrence is relentless.
5. During recurrence, metastases of the regional lymph nodes and occasionally of the viscera and skeleton may occur.
6. These tumours are radioresistant.
7. Excision of these tumours is better followed by radiotherapy.

SJOGREN'S DISEASE

It is regarded as an *autoimmune disorder*—the antibodies arise in response to antigen derived from the nuclei and cytoplasm of the cells of the parotid gland. It involves the salivary glands, lacrimal glands, mucous glands and is *associated with some other systemic manifestations*.

Histology reveals replacement of salivary tissue by lymphoid tissue containing islets of epithelial tissue.

Some pathologists consider this condition to be due to an obscure chronic inflammation.

Characteristic features

1. Commonly found in women above the age of 40 years.
2. The condition is usually bilateral.
3. There will be symmetrical enlargement of the salivary glands (*commonly parotid glands*).
4. Involvement of the *lacrimal glands* causing narrowing of the palpebral fissures.
5. Dryness of the mouth and eyes.



Fig. 5.12. Mikulicz disease. Note bilateral enlarged submandibular salivary glands and bilateral enlarged lacrimal glands causing narrowing of the palpable fissures. (Courtesy: Prof. Tarun Chatterjee, MS)

When associated with any systemic manifestations like rheumatoid arthritis the condition is addressed as Sjogren's disease.

6. *Systemic manifestations:* There will be generalised arthritis particularly of the small joints (rheumatoid type), keratoconjunctivitis, scleroderma, polyarteritis nodosa.
7. The condition develops slowly and painlessly.

Plain X-ray occasionally reveals small areas of calcification throughout the involved gland.

MIKULICZ DISEASE

It is also an autoimmune disorder and is regarded as a variety of Sjogren's disease *where all the salivary and lacrimal glands are enlarged, but without any systemic manifestation.*

Treatment

1. Spontaneous cure may occur after several weeks or several years.
2. Steroid therapy may be of help.
3. Radiotherapy may be employed when the diagnosis is confirmed.
4. Surgery: If there are cases of massive deformity then removal of submandibular salivary glands and bilateral superficial lobectomy of the parotids are advocated. *The lacrimal glands are better left alone.*

The surgery should be followed by either steroid therapy or radiotherapy.

6

CHAPTER



Swellings of the Jaw

Swellings of the jaw are caused by innumerable lesions and they may arise from the mucoperiosteum or from the tooth-germs or from the bones.

I. Arising from the mucoperiosteum, i.e., epulis

Epulis — May be:

1. Fibrous
2. Granulomatous
3. Myeloid
4. Sarcomatous
5. Carcinomatous.

II. Arising from the tooth germs, i.e., odontomes (after McGregor)

Odontomes — May be:

1. Epithelial odontomes, i.e., arising from epithelial elements:
 - Dental cyst
 - Dentigerous cyst
 - Adamantinoma.
2. Mesothelial odontomes, i.e., arising from connective tissue elements:
 - Fibrous odontomes
 - Cementomes
 - Osseous or sarcomatous odontomes
3. Composite odontomes, i.e., arising from both epithelial and mesothelial elements:
 - Radicular odontomes
 - Osseous or sarcomatous odontomes

Rare

Extremely rare

III. Arising from the bones

1. Traumatic
2. Inflammatory
 - Specific, e.g., actinomycosis
 - Non-specific, e.g., osteomyelitis, alveolar abscess
3. Neoplastic
 - Benign
 - Malignant.

Benign:

- (a) Arising from fibrous tissue — fibrous dysplasia.
- (b) Arising from osteoblasts —
 - Osteoma — localised or diffused
 - Osteoclastoma.

Malignant:

- (a) Primary — carcinoma and sarcoma.
- (b) Secondary, i.e., by extension from an adjacent growth, e.g., carcinoma of the tongue involving the lower jaw.

EPULIS

The word 'epulis' means upon the gum. It is a solid swelling situated on the gum and arises from the alveolar margin of the jaw. *It can originate from the mucous membrane, periosteum, or bone.* Usually it occurs secondary to chronic inflammation and sometimes it is considered as hamartoma.

Following varieties of epulis are commonly encountered:

- | | |
|------------------|-------------|
| 1. Fibrous | } Benign |
| 2. Granulomatous | |
| 3. Myeloid | |
| 4. Sarcomatous | } Malignant |
| 5. Carcinomatous | |

FIBROUS EPULIS

Commonest variety. It arises from the periosteum or from the periodontal membrane and forms a firm nodule at the junction of the gum and tooth. Chronic dental sepsis is considered to be an important predisposing factor.

Histologically, it is whorled fibroma, and is composed of fusiform cells with many new blood vessels.

Diagnostic criteria

1. More common in women and in young adults.
2. Lower gum is commonly affected.
3. More commonly found on the external aspect of the gum, *at the neck of canine or premolar tooth*.
4. It may be sessile or pedunculated.
5. It is a slowly growing, firm swelling and not tender.
6. Overlying mucous membrane is healthy but it is grey or slightly brighter pink than that of the normal gum.
7. The adjacent teeth are irregularly displaced and loosened and the tumour may protrude between the irregular teeth.
8. Draining lymph nodes are not enlarged.

Complications

1. Although it is benign, it has a tendency to recur after operation, if its root is not thoroughly excised.
2. Malignant change—fibrosarcoma, rare.

Treatment

Excision of the swelling. *Adjacent tooth and a wedge of bone around its root may be removed to prevent recurrence.*

GRANULOMATOUS EPULIS

It consists of a mass of granulation tissue which forms *around a carious tooth or at the site of chronic*

irritation by a denture. This is not a true epulis and hence considered false epulis.

Sometimes, a similar condition may be found temporarily during pregnancy which is known as gingivitis gravidarum.

Diagnostic criteria

1. Rapidly growing soft swelling, bright red in colour.
2. The overlying mucous membrane is granular, persistently ulcerated. It bleeds on touch or during brushing of the teeth.
3. Offensive smell due to infection.
4. Presence of adjacent carious tooth or ill-fitting denture.
5. Draining lymph nodes are enlarged and tender.

Treatment

1. Scraping of the granulation tissue.
2. Extraction of the carious tooth.
3. Pre- and postoperative antibiotics to combat infection.
4. Maintenance of oral hygiene.
5. Any ill-fitting denture should be replaced.

MYELOID EPULIS

Syn.: Giant cell epulis

Pathologically, *it is an osteoclastoma*, arising from the mucoperiosteum of the gum.

Histologically, it consists of fibrocellular tissue scattered through which are multinucleated giant cells.

Diagnostic criteria

1. Commonly occurs between the ages of 10 and 25 years.
2. *Common in the lower jaw, near the angle of the mandible.*
3. It is a rapidly growing painless tumour presenting under the gum.
4. The swelling is soft and sessile, surface is smooth and lobulated. The overlying mucous membrane of the gum becomes hyperaemic and oedematous. It looks dark-red or plum-coloured and may bleed and ulcerate.
5. *Expansion of the bone occurs—both externally and internally (cf. Adamantinoma).*
6. In a well developed case there may be the presence of egg-shell cracking.

7. Draining lymph nodes are not enlarged.
8. Confirmation by X-ray – soap-bubble or honey-comb appearance due to bone destruction with ridging of the walls (pseudotrabeulation).

Complications

1. Ulceration.
2. Haemorrhage which may be severe.

Treatment

1. *If the swelling is small* – curettage of the tumour followed by cauterisation and filling up of the remaining cavity by cancellous bone chips taken from the iliac crest.
2. *When the swelling is of a big size* – radical excision of the tumour with adjacent margin of healthy bone followed by grafting for the mandible. The graft is taken usually from a rib.

CARCINOMATOUS EPULIS

It is an *epithelioma of the gum* arising from the mucosa of the alveolar margin. It may start around a tooth, or in a socket, giving rise to a painful, infected and infiltrating lesion. Later on the lesion becomes frankly fungating and ulcerating and invades the bone. Cervical lymph nodes are enlarged.

Biopsy should be performed to confirm the diagnosis.

Treatment

1. Radical resection followed by grafting for the mandible. When it occurs in relation to upper jaw, adequate resection of the maxilla should be done.
2. Radiotherapy – may be of help in selected cases.

ODONTOMES

Odontomes are cysts, tooth malformations, or tumours arising from the tooth germs. *Truly they are malformations but not tumours* (Willis). They usually arise from the epithelial or mesothelial elements of the teeth, or sometimes from both.

In human beings epithelial odontomes are commonly encountered and there are three important epithelial odontomes, namely:

1. Dental cyst
2. Dentigerous cyst
3. Adamantinoma.

Basis of origin of epithelial odontomes

During development of tooth there occurs an ingrowth of epithelium (ectoderm) lining the mouth over the site of the future gums. *This epithelial element later forms the enamel organ of tooth and is called ameloblast.* Clusters of this epithelium may persist as epithelial debris or epithelial rest and later may grow into cysts or tumours.

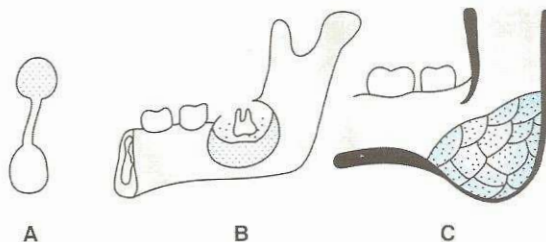


Fig. 6.1. Common types of odontomes. (A) dental cyst; (B) dentigerous cyst; (C) adamantinoma.

DENTAL CYST

Syn.: Radicular cyst, Periodontal cyst or Epitheliated granuloma

This is a unilocular cyst arising in connection with the root of a normally erupted, but chronically infected and usually pulpless tooth (Fig. 6.1).

The continued irritation of infection appears to stimulate the epithelial cells, believed to be derived from the enamel organ. As a result these epithelial cells proliferate and degenerate to form a cyst.

Pathology

Lining of the cyst wall: Squamous epithelium.

Contents: The contents of the cyst may be fluid or semisolid in nature, containing cellular debris with cholesterol crystals and foreign body giant cells. As a rule, the content is sterile on culture but secondary infection can occur.

The cyst enlarges slowly causing expansion of the alveolus, and eventually most of the epithelial lining is destroyed, if infection remains active.

Diagnostic criteria

1. It can appear at any age, but commonly in or after middle age.
2. Usually painless swelling unless secondarily infected.

3. It commonly affects the upper jaw, in association with a carious tooth or pulpless tooth – incisor or canine tooth.
4. It causes an expansion of the bone as it grows, and in advanced cases there may be egg-shell crackling.
5. There may be fluctuation, if the bone is completely destroyed.

X-ray: An oval or circular radio-translucent area in relation to the root of a tooth. The margin of the translucent area is clear but sclerosed.

DENTIGEROUS CYST

Syn.: Follicular odontome or cyst

This is a unilocular cyst arising in *connection with a non-erupted but permanent tooth* (cf. Dental cyst). The lesion arises from the follicle of the developing tooth. The follicle consists of an inner epithelial lining and an outer connective tissue covering.

Pathology

Lining of the cyst wall: Usually fibrous, but may be lined by squamous cell epithelium derived from the enamel producing cells.

The crown of the tooth inside the cyst lacks the dental cuticle.

Contents: The contents of the cyst may be glairy and viscid fluid or semisolid in nature, with cholesterol crystals and giant cells. It also contains unerupted tooth, commonly an upper or lower third molar tooth, with truncated root. The tooth lies either free in the cavity lying obliquely or, rarely, the tooth lies embedded in the wall of the cyst (Fig. 6.1).

Diagnostic criteria

1. Any age may be affected but more common in teens.
2. A painless, slowly growing swelling at the site of an unerupted tooth.
3. More commonly occurring in the lower jaw in the third molar tooth region.
4. The tooth is missing at the site of a dentigerous cyst (but on counting during examination the tooth may be found normal, because the milk tooth may persist).
5. There is an expansion of the outer table of the jaw even causing egg-shell crackling. The inner table remains strong and prevents pathological fracture.

X-ray or orthopantomogram: A well circumscribed radio-translucent area with the crown of the tooth inside it. Similar X-ray findings may be noticed in other lesion of the jaw like adenomatoid odontogenic tumour. There may be pseudotrabeulation or soap-bubble appearance due to ridges of bone on the side walls.

Treatment of dental and dentigerous cyst Operative

1. Total excision of the cyst wall. The steps are:
Intra-oral approach: Two types of incision are in use:
 - (a) A transversely placed curved incision is made over the mucoperiosteum at the maximum point of convexity of the swelling. Then the mucoperiosteal flaps are raised.
 - (b) Three-sided flap incision with broader base at the mucobuccal junction is made to raise the mucoperiosteal flap.
 - An opening is made in the thin outer shell of the bone; the cyst is entered.
 - The cyst is thoroughly scooped out along with its whole epithelial lining and the related tooth.
 - After thorough excision, the bone cavity is obliterated by soft tissue 'push-in' or with the bone chips.
 - The mucoperiosteal flap is approximated and sutured.
2. If the cyst is too big and total excision is not possible, then the cyst is de-roofed, scooped out as much as possible and marsupialized.
3. Any carious tooth should be removed.
4. Pre- and postoperative dental hygiene, and control of infection.

ADAMANTINOMA

Syn.: Ameloblastoma; Multicystic disease of the jaw; Fibrocystic disease of the jaw; Eve's disease

Origin of the lesion

Regarding its origin there are two views:

1. It is an epithelial tumour arising from the ameloblast which is the primitive enamel forming cells of the tooth.
2. According to Willis, it is a basal-cell carcinoma arising from the ectodermal epithelium of the

stomodeum. It behaves pathologically and looks histologically like a basal-celled carcinoma. The histological picture and recurrence of the tumour after incomplete removal supports the view of basal cell carcinoma.

Pathology

It is a painless slowly growing, benign, solid tumour. Subsequently it undergoes multi-centric cystic degeneration, causing simultaneous resorption of the bone and expansion of the jaw – mostly of the outer table in the region of molar tooth, with thinning of the cortex and deposition of periosteal new bone. Some of the loculi are separated by bony trabeculae.

Although benign, it may infiltrate the bone further than what is apparent to the naked eye, so that *removal of the tumour by curettage may be followed by recurrence. But it does not metastasize to lymph nodes.*

Lining of the cyst wall: Either squamous or columnar cell epithelium with areas of fibrous and osseous tissue.

Contents: Yellowish mucoid material. Very rarely adamantinoma may contain melanin pigment.

Histological picture

Usually it consists of an outer layer of deeply basophilic columnar cells – the *enameloblasts*, and a central core of 'Star-cells', i.e., cells with large vacuoles in the cytoplasm and connecting cytoplasmic bridges.

In other cases, there is presence of sheets of epidermoid cells resembling a basal cell carcinoma, but

presence of isolated clumps of columnar *enameloblasts* will usually indicate the true nature of the lesion.

Diagnostic criteria

1. Age: Majority of the tumours occur between 10 and 35 years, and after 50 years they are rare.
2. Sex: Females are commonly affected.
3. It is a slowly growing, painless tumour.
4. Commonly affecting the mandible in the region of the molar teeth (i.e., near the angle of the jaw).
5. It causes expansion with thinning out mostly of the outer table (cf. osteoclastoma) and so gives rise to a swelling more often on the cheek than within the mouth. With a marked expansion of the bone, there may be egg-shell crackling on palpation.
6. Occasionally due to gross cystic degeneration, the tumour may be soft and fluctuant.
7. Generally there is no missing tooth. There may be malalignment and malocclusion of the neighbouring teeth.
8. The tumour may attain a large size to produce an ugly deformity of the face.
9. Cervical lymph nodes are not enlarged.

X-ray or orthopantomogram: Soap-bubble appearance, or honey-comb appearance, i.e., multiple small translucent areas separated by well-formed bony trabeculae (Fig. 6.1C). There is expansion of the outer table of the mandible. Skiagraphy is very often simulated with that of osteoclastoma.

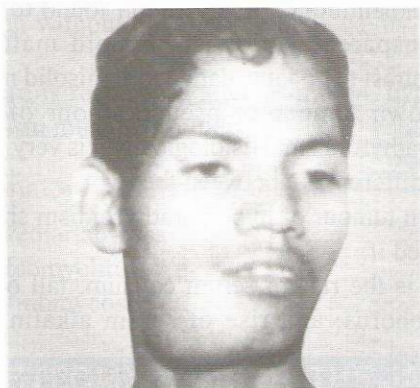


Fig. 6.2. Adamantinoma lower jaw – a slowly growing, painless tumour involving the lower end of mandible near the angle of the jaw.

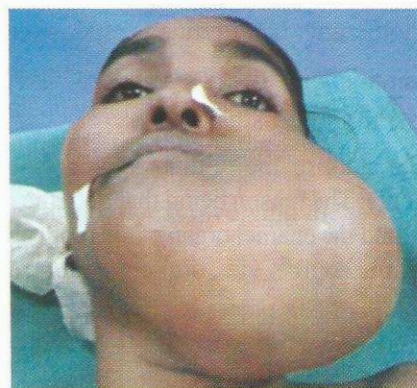


Fig. 6.3. Large-sized adamantoma of the left lower jaw. (Courtesy: Prof. R.N. Poddar, R. Ahmed Dental College, Kolkata)

Differential diagnosis

1. Osteoclastoma or giant cell tumour.
2. Giant cell reparative granuloma of the jaw.

Treatment

1. *Evacuation and curettage of the cyst* with or without bone grafting: There is every chance of recurrence, though recurrence does not cause any lymph node metastasis.
2. *Selective block excision of the bone along with tumour*: If the basal bone of the mandible is not affected by the tumour, the tumour is excised with a generous amount of surrounding healthy bone leaving the lower border of the mandible intact to maintain the continuity of the jaw. The remaining cavity is electrocauterised and may be filled with marrow cancellous bone graft.
3. *Partial resection of the segment of the jaw bearing the tumour*, together with a margin of surrounding healthy bone.
4. *If the tumour is too large in size, affecting the ramus of the mandible, or is of advanced stage, hemimandibulectomy is necessary*. Following resection of the segment of the jaw or hemimandibulectomy, the defect can be filled up by an immediate bone graft. Failure of immediate bone graft is substituted by a holding prosthesis such as that of Bowerman & Conroy, or a silastic rod carved to the design and skewed over a K-wire. After a period of 3 to 6 months the holding prosthesis is dissected and replaced by a block bone graft, or marrow cancellous bone graft put in a tray of tantalum mesh bone implant.
5. Radiotherapy: Not of much value.

Can adamantinoma occur elsewhere than the jaw?

Yes, in the following situations:

1. In the stalk of the pituitary gland (suprasellar tumour): The reason being that both the pituitary stalk and the enamel organ of the tooth arise from the same source, viz., oral epithelium.
2. In the tibia: The exact reason is not known. There are three hypothesis:
 - (a) Abnormal embryonic inclusion of tooth-germ epithelium within the developing tibia giving rise to neoplasm many years later.

- (b) Traumatic separation of the epithelium of the skin and implantation beneath the periosteum, even in the absence of any history of a penetrating wound, though history of trauma is often present.
- (c) The lesion may be an apparent atypical synovioma.

GIANT-CELLED REPARATIVE GRANULOMA

This is a condition arising due to occurrence of haemorrhage within the bone marrow causing an expansion of the bony wall.

Pathology

- *Naked eye appearance*: During operation found to be consisting of opaque, semisolid, dark-red material.
- *Histologically*, consisting of small multinuclear cells which are sparsely and unevenly distributed.

Diagnostic criteria

1. Age: Nearly always between the ages of 10 and 25 years.
2. More common in females.
3. A painless swelling of the jaw, situated more frequently in the mandible.

X-ray: Rounded or oval area of translucency with expansion and thinning out of the cortex.

Differential diagnosis

1. Osteoclastoma: By age, osteoclastoma appears in patients between 25 and 40 years of age.
2. Adamantinoma: At operation found to contain transparent yellowish mucoid material in contrast to opaque, dark-red semisolid material.
3. Brown tumour or pseudotumour of hyperparathyroidism: Histologically it is very difficult to differentiate between the two.

Brown tumour of hyperparathyroidism should be suspected if:

- there is the rise of serum calcium, fall of serum phosphorus, and rise of serum alkaline phosphatase;
- X-ray of other parts of the skeleton will show subperiosteal bone resorption giving rise to small clear cystic spaces.

Differential diagnosis of osteoclastoma, adamantinoma and giant-cell reparative granuloma affecting the jaw

	Osteoclastoma	Adamantinoma	Giant-cell reparative granuloma
<i>Frequency</i>	Less common	More common	Less common
<i>Age</i>	Commonly between 25 and 40 years	Commonly between 10 and 35 years	Between 10 and 25 years
<i>Sex</i>	Males – more common	Females – more common	Females – more common
<i>Rate of growth</i>	Relatively rapidly growing	Slowly growing	Slowly growing
<i>Expansion of bony walls</i>	Both the inner and outer tables are expanded	Outer table is expanded	Both the tables are expanded
<i>Egg-shell crackling</i>	More frequent	Less frequent	Frequent
<i>Pathological fracture</i>	Not uncommon	Very rare	Rare
<i>Fungation</i>	May occur	Very unusual	Very unusual
<i>Sinus/fistula</i>	Rare	May occur externally or internally	Rare
<i>Radiography</i>	Soap-bubble or honeycomb appearance – the cysts are larger in size with fine and ill-defined trabeculations (pseudo-trabeculations)	Same appearance but the cysts are smaller in size with well-marked trabeculations	Rounded or oval translucent cystic area with or without faint trabeculations
<i>Histology</i>	Features of osteoclastoma – presence of multinucleated giant cells in fibrocellular stroma	Presence of peripheral columnar cells – the enameloblasts, with a central core of star cells	Presence of small multinucleated cells which are sparsely and unevenly distributed
<i>Radio-sensitivity</i>	Radiosensitive	Non-radiosensitive	Does not arise
<i>Recurrence</i>	Common	Common – particularly when evacuation and curettage is done	Nil. If it at all occurs, hyperparathyroidism should be suspected and investigated
<i>Malignancy</i>	May occur in 10 percent cases	Very uncommon	Nil

Treatment

Thorough curettage of the cavity through external approach without opening the bone cavity into the mouth.

If there is recurrence after operation, hyperparathyroidism must be excluded.

Inflammatory swelling of the jaw

1. Alveolar abscess
2. Osteomyelitis
3. Actinomycosis

OSTEOMYELITIS OF THE JAW

Syn.: Necrosis of the jaw

This may be:

1. Acute – very rare.

2. Subacute.
3. Chronic.

Predisposing factors

1. Spread of apical dental infection.
2. Inadequate drainage of an alveolar abscess.
3. Compound fracture of the jaw.
4. Radiation necrosis.
5. Buccal carcinoma involving the jaw.
6. Cancrum oris.
7. Chemical:
 - Phosphate (leading to phossy jaw/necrosis)
 - Arsenic
 - Mercury.
8. Actinomycosis, syphilis and tuberculosis rarely.

Organism

Staphylococcus aureus.

Which jaw is commonly affected?

The lower jaw, i.e., the mandible is commonly affected, because it has a single, rather tenuous blood supply (inferior alveolar artery) running along the long axis of the bone. Therefore, it is easily obstructed by infection or trauma, leading to necrosis of the bone, which is the essence of osteomyelitis of the jaw.

The upper jaw, i.e., the maxilla is less commonly affected, because the blood supply of the upper jaw is segmental and vertical with anastomoses with each other. Therefore, it is not dependent on a single vessel and thus infection or trauma is most unlikely to cause complete cutting off of the blood supply to the bone.



Fig. 6.4. Chronic suppurative osteomyelitis of the mandible. (Courtesy: Dr. S. Jayachandran, MDS)

Diagnostic criteria

1. Commonly, the mandible is affected.
2. History of any predisposing factors, e.g., dental extraction or incompletely treated alveolar abscess, etc.
3. Insidious onset of a painful swelling with or without fever. The pain is due to tension within the bone.
4. Presence of either internal or external discharging sinus – may be due to spontaneous bursting which may relieve tension within the bone, and so pain is relieved on discharge of pus. The sinus is usually fixed to the underlying bone.
5. History of extrusion of bone chips may be there.

6. On palpation: The bone thickening is not a prominent feature, because reparative bone formation is poor (as opposed to osteomyelitis of the other sites).
7. The swelling is tender.
8. The draining lymph nodes are enlarged and tender.
9. Paraesthesia or numbness of the chin in the distribution of the mental nerve may be present (because increased tension within the dental canal compresses the inferior alveolar nerve as well as the vessels).

X-ray: One of the following pictures is evident:

- Evidence of local osteitis with periosteal reaction.
- A small cavity (due to bone destruction) with surrounding sclerosis with or without a sequestrum. The sequestrum formation is late as the jaw bones are developed from membrane. Sometimes the sequestrum may be a massive one and the lower jaw may collapse leading to an ugly deformity of the face.

Occasionally, the offending broken root of a tooth may be seen within the cavity.

Treatment

1. When osteomyelitis is suspected clinically but not confirmed radiologically, X-rays must be taken at regular intervals (weekly). As soon as the bone necrosis will be evident, operation should be done to relieve tension and remove the diseased materials. *Radiological evidence of bone necrosis takes at least 3 weeks to become manifest.*
2. If there is a discharging sinus, pus should be sent for culture and sensitivity to antibiotics.
3. Both pre- and postoperative suitable antibiotics should be given according to sensitivity test.
4. In a well proved case, dependent drainage gives the best result.

Steps of operation:

- Incision is made about 1/4" below the lower border of mandible. It should not extend beyond the point one inch below and in front of the angle of the mandible (to avoid injury to the mandibular division of the facial nerve,

otherwise there will be paralysis of the lower lip).

- Periosteum is elevated.
 - A trough is chiseled in the bone carefully to avoid fracture of the jaw.
 - Any unhealthy granulation tissue should be scraped thoroughly.
 - Sequestrum, if present, is removed.
 - The sclerosed cavity should be deroofed.
- Skin may be closed, or the wound may be left open and packed lightly with sterile petroleum jelly gauze.
5. Infected tooth, if any remaining, should be extracted.

ACTINOMYCOSIS OF THE JAW

Vide actinomycosis (Chapter 7).

7

CHAPTER



Actinomycosis

Actinomycosis is a chronic, suppurative granulomatous disease occurring particularly in the region of the lower jaw. The casual organism is potentially pathogenic fungus-like organism *Actinomyces israelii*.

Bacteriology: *A. israelii* is an *anaerobic gram-positive organism*. This organism is now considered to be a bacterium in the family Actinomycetaceae which also includes the genus *Nocardia*; the *Actinomyces* is non-acid-fast and *Nocardia* is relatively acid-fast.

In the tissues, the *Actinomyces* grow in the form of complete colonies which can be recognised in the pus by the naked eye as pinhead granules, greyish yellow in colour. These are called 'sulphur granules' or 'fish-roe' bodies. When teased out on a microscopic slide and examined unstained they are seen to consist of branching mycelial filaments. Each filament ends in a terminal club.

The clubs are gram-negative, and are not formed in artificial culture. It is believed that they are produced as the result of deposition of the colony of lipid material derived from the host tissues. The filaments are gram-positive. These organisms are found in the colonies in characteristic radial arrangement and hence popularly known as 'ray-fungus'.

Natural habitat: *A. israelii* is a normal commensal of the mouth and it is found in the tonsils and carious

teeth and infected gum without any clinical manifestations of the disease. *It may also be found in the large intestines.*

Mode of infection: Actinomycosis is an *endogenous infection*. It gains access to the tissue planes through the crypts or defects in the mucous membrane as a result of ulceration or trauma (e.g., following dental extraction).

The actinomycotic lesion starts as an acute suppurative inflammation which ultimately progresses to chronicity. A granulomatous mass is produced and it comprises multiple nodules of fibrocellular tissue. Some of the nodules liquefy to form multiple small chronic abscesses. Some of them fuse together but others remain discrete owing to the persistence of fibrous septa. This produces a characteristically loculated appearance (honeycomb appearance).

Some of the abscesses burst to the surface to discharge necrotic yellowish material through multiple sinuses. Thus a hard brawny painless swelling with multiple discharging sinuses is developed.

Clinical types of actinomycosis

Cervicofacial	65 percent
Ileocaecal	25 percent
Pulmonary	10 percent

Spread

Infection spreads by continuity and contiguity. In cervicofacial actinomycosis there is a direct spread

to the adjacent connective tissue, the muscle and the bone which are replaced by granulation tissue with an abundance of fibrous tissue reaction. There is also progressive cutaneous involvement resulting in *multiple discharging sinuses*.

Lymphatic spread does not occur in actinomycosis, because the filaments cannot pass through the lymphatic channels. Any regional lymphadenitis, if it occurs, is due to secondary bacterial infection.

Blood spread may occur. Liver may be involved either via the hepatic artery from cervicofacial lesion, or via the portal vein from ileocaecal lesion. Pulmonary actinomycosis may disseminate into the systemic circulation and thus produce lesion in the kidney, brain, bone and other parts.

Prognosis

The prognosis of cervicofacial actinomycosis is good, but the abdominal and pulmonary infections are sometimes fatal.

Punch actinomycosis

Rarely there may be actinomycotic infection in a hand wound caused by hitting an assailant in the teeth.

CERVICOFACIAL (NECK) ACTINOMYCOSIS

Diagnostic criteria

1. Common site – in the region of the lower jaw.
2. Very slowly growing.
3. *A hard brawny indurated painless swelling with multiple discharging sinuses.*
4. Bluish discolouration of the overlying skin.
5. Characteristic *yellow sulphur granules* can be demonstrated in the discharging sinuses.
6. X-ray may reveal multiple sclerosed bony cavities with sequestra in the lower jaw.
7. On microscopic examination – Granules consist of branching mycelial filaments with terminal clubs.

Treatment

1. Medical treatment
2. Surgical treatment
3. Radiotherapy.

Medical treatment

Actinomycin is usually sensitive to penicillin, tetracycline, lincomycin.

A massive dose of penicillin – to start with 10 mega units, then reducing it to 4 mega units I.M. daily – is administered for a prolonged period and the treatment should be continued at least for 3 months after clinical cure.

In resistant cases broad-spectrum antibiotics should be tried after antibiotic sensitivity test.

Iodides: The penicillin therapy is supplemented with oral administration of tincture iodine in the dose of m X to XV in a glass of milk 3 times a day. *The iodides are supposed to help in the resolution of fibrosis.*

Surgical treatment

Incision and drainage: Under the cover of antibiotic the abscesses and sinuses should better be opened up and the wound is then loosely packed up with roller gauze soaked in tincture iodine (2 percent). Excision of the granulomatous tissue is very difficult.

If there is osteomyelitis of the jaw, the bony cavity should be drained and sequestra, when present, be removed.

Radiotherapy

In resistant cases radiotherapy may be tried.

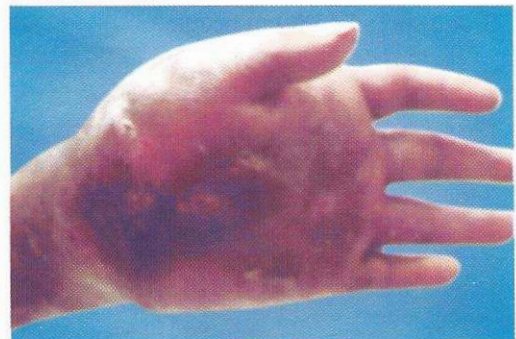


Fig. 7.1. Actinomycosis of the left hand. A hard brawny indurated painless swelling with multiple discharging sinuses. The discharge was yellowish in colour. (Courtesy: Dr. Yogesh Salphale)

MADURA FOOT (MYCETOMA PEDIS)

It is a chronic infective granulomatous condition affecting the subcutaneous tissues of the foot caused

by the organism known as 'aerobic streptothrix'. There are two types of organisms:

1. Bacterial known as *Actinomycosis nocardia*.
2. Fungal known as *Madura mycosis*.

Nocardia infection is common. *Nocardia* are aerobic, gram-positive, and often acid-fast.

These organisms are usually found in the soil and they are introduced into the skin and subcutaneous tissue through minute abrasions caused by trivial injury – so, *exogenous infection* (cf. *Actinomycosis israelii*).

The condition is common in tropical countries, and is endemic in Madura district of South India, hence it is named *Madura foot*. Most of the patients are farmers who walk barefooted. The condition may also occur in other parts of the body besides the foot.

In the early stage, pale, firm nodules appear. The nodules are surrounded with vesicles. Some of the nodules liquefy and result in chronic abscess formation. Some of the abscess burst and multiple discharging sinuses develop over the top of the nodules. The discharging granules may be yellow, black or red in colour. In the late stage, there will be invasion of deeper structures like tendon, muscles, and bone.

Spread

1. Spreads by direct continuity and contiguity.
2. Lymphatic spread does not occur.
3. Blood spread does not occur.



Fig. 7.2. A hard, brawny, indurated swelling with multiple discharging sinuses in the left foot of a farmer. The discharge was black in colour — Madura foot. D/D: Koch's osteomyelitis.

Diagnostic criteria

1. Common in farmers who walk bare-footed.
2. Common in southern part of India.
3. A chronic, firm to hard, brawny, indurated painless swelling with multiple discharging sinuses. The foot is oedematous.
4. The overlying skin shows bluish discolouration.
5. Discharging granules may be yellow, black or red in colour.
6. In the late stage, the swelling may be fixed to the underlying bone, and becomes thick and tender.
7. X-ray: In *actinomycosis Nocardia* type, X-ray reveals erosion of bones with sclerosis but no sequestra.
In *Madura mycosis* type X-ray reveals multiple rarefied circular areas in the bones looking like a honeycomb.
8. Microscopic examination of the granules and biopsy reveal characteristic organisms.

Treatment

1. Medical treatment
2. Surgical treatment

Medical treatment

1. All cases should be treated with massive dosage of penicillin.
2. Dapsone (diaminodiphenylsulphone) — Sulphone group of drugs are useful in *Nocardia* infection (bacterial), but are of little or no value against *Madura mycotic* type (fungal). Dose is 100 mg twice a day for a period of 2 years.
3. Oral iodides may be given to help the resolution of fibrosis.

Surgical treatment

1. Multiple incisions and drainage under the cover of penicillin and dapsone.
2. In resistant cases, amputation is required at a suitable level, which depends on the site and spread of the disease.

Prognosis

The prognosis is poor and unfortunately the disease recurs in about 20 to 30 percent of cases.

8

CHAPTER

The Mouth and the Tongue



CYSTS IN THE MOUTH

The following cysts may be encountered:

1. Mucous cysts.
2. Dermoid, see Chapter 2.
3. Ranula, see Chapter 2.

MUCOUS CYSTS

The inner surface of the lips and the cheek is covered with an epithelium that contains many tiny mucus-secreting glands. Obstruction of the duct of one of the glands causes *mucus-retention cyst*. Often mucus escapes into the tissues following rupture of the duct and causes mucus-extravasation cyst.

Diagnostic criteria

1. Age: These cysts can occur at all ages.
2. The patient complains of a slowly growing lump on the inside of the lips or cheek. The lump may interfere with eating and get bitten through.
3. Site: *Commonly present on the lower lip*. It may occur on the buccal mucous membrane.
4. Spherical or globular in shape, and the size varies from 0.5 to 2 cm in diameter.
5. *Translucent, soft cystic swelling* having a pale pink colour with a smooth surface. The overlying epithelium may be white and scarred if the cyst has been frequently damaged by biting.



Fig. 8.1. Mucous cyst – lower lip.

Complications

1. Spontaneous rupture
2. Recurrence
3. Gets bitten through.

Treatment

1. Excision under local anaesthesia.
2. Injections of boiling water when the cysts are multiple.
3. Cryosurgery.

TUMOURS OF THE CHEEK AND FLOOR OF THE MOUTH

Papilloma

1. It is usually uncommon.
2. It appears as a soft, fleshy mass.
3. Chance of malignancy is remote.

4. Treatment: Excision with an adequate margin of tissue followed by histopathology examination of the excised tissue.

Fibroepithelial polyp

1. It appears as a firm pale swelling on the inner surface of the cheek.
2. It arises after repeated trauma to mucous membrane of the cheek.
3. Histologically, it contains a dense core of fibrous tissue.
4. Treated by excision.

Lipoma

It can arise in any part of the fatty supporting structures of the mouth. Features are the same as with other lipomas. Treated by excision.

Haemangioma

It can arise under the mucous membrane of the cheek and floor of the mouth, lips and tongue. Features are the same as with other haemangiomas. Recurrent bleeding after getting bitten through is common.

Treatment

- Injection of boiling water.
- Cryosurgery.
- Excision.

Ectopic salivary tumours

See page 129.

LEUKOPLAKIA

This is a descriptive term meaning *white patches* on a squamous epithelium in or near a mucous surface. This condition may occur anywhere within the mouth, commonly on the tongue. *It is basically an increased keratinising process (hyperkeratosis) of the superficial cells of the squamous epithelium which occurs in response to a chronic irritant, e.g., sharp tooth or ill-fitting dentures, smoking, sepsis, spirits and spices, syphilis (well known list of S's).*

Other sites are the larynx, the perianal region, glans penis, and the vulva.

Macroscopically, the affected area of mucous membrane appears as a *thickened grey-white plaque* with/without cracks or fissures. The plaque is difficult to rub-off.

Microscopically, there is hyperplasia of superficial layers of squamous epithelium with hyperkeratosis,

swelling and vacuolation of cells of the middle layer, and hyperplasia and hyperchromasia of the basal layers of cells. There may be dyskeratosis which is most serious because of its association with malignancy.

The importance of the lesion is that *it is pre-malignant* and the frequency of the neoplastic change is about 25 percent. The neoplastic change usually occurs within the fissures and should be suspected if there is local thickening, bleeding, area of erythema, appearance of nodules, induration around the white patch, or pain.

Diagnostic criteria

1. Age: Usually found in middle aged or elderly subjects.
2. Sex: Men are more affected.
3. Patient may present with one of the following features which are divided into 4 clinical stages:

- | | |
|-----------|---|
| Stage I | Appearance of thin grey transparent patch on the tongue which may be localised or widespread. |
| Stage II | This thin patch gradually turns white and opaque. This is leukoplakia. In the beginning it looks soft but later cracks and fissures appear. |
| Stage III | Gradually hyperplasia will lead to small nodules or warty excrescences. Desquamation may follow which leaves areas of smooth, red and shiny patches. |
| Stage IV | This is the stage of appearance of carcinoma. The lesion possesses all the characteristic features of carcinoma. <i>The carcinomatous change usually occurs within the fissures and should be suspected, if there is local thickening, bleeding, or pain.</i> |

Treatment

1. Removal of any underlying cause, e.g., removal of the sharp tooth causing chronic irritation, care of oral hygiene. Smoking, chewing tobacco and alcohol should be stopped.
2. Any indurated white patch suspicious of malignancy should be biopsied at once.
3. (a) Small superficial patches of leukoplakia may be satisfactorily treated by excision.
(b) Larger lesion requires excision followed by closure with mobilisation of local flaps or skin grafting. Rarely, the raw area after excision is left for epithelialisation.

4. Patients should be examined at regular intervals of not longer than three months to note the appearance of any area of warty excrescence or any doubtful area which must be removed and examined histologically.
5. Radiotherapy: Its role is intricate. Although it improves the condition initially, it increases the chance of malignancy. Moreover such malignant lesions become resistant to further radiotherapy.

ULCERS OF THE TONGUE

Types

Non-specific

1. Dyspeptic or Aphthous ulcer
2. Traumatic or Dental ulcer
3. Infective and allied:
 - Chronic non-specific ulcer
 - Post-pertussis ulcer
 - Simple ulcer due to glossitis
 - Herpetic and pseudo-herpetic ulcer.

Specific

1. Tubercular ulcer
2. Syphilitic ulcer
3. Malignant ulcer

Differential diagnosis of tongue ulcers

See page 151.

Management of tongue ulcer in elderly persons

After thorough clinical examination and investigation, *biopsy of the ulcer is a must* in every case and the tissue should be sent for histopathological examination to exclude carcinoma.

TUMOURS OF THE TONGUE

Benign and malignant

Benign tumours of the tongue are very uncommon. The following may be encountered rarely.

- Papilloma – Sessile or pedunculated
- Haemangioma – Usually cavernous type
- Lymphangioma
- Neurofibroma, plexiform neuroma
- Lipoma
- Lingual thyroid
- Mixed salivary gland tumour
- Osteoma

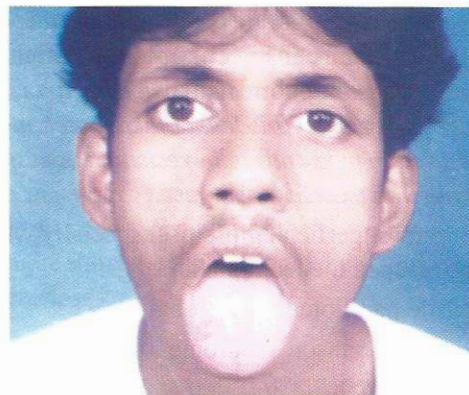


Fig. 8.2. Papilloma of the tongue. Treatment: excision biopsy.

Malignant tumours

- Carcinoma
- Sarcoma.

MACROGLOSSIA

Macroglossia is a large painless tongue. The causes are:

1. Lymphangioma
2. Haemangioma
3. Neurofibroma
4. Primary mesodermal amyloidosis
5. Muscle hypertrophy (seen in cretins)
6. Myxoedema.

CARCINOMA OF THE TONGUE

The squamous-cell mucous membrane lining the buccal cavity, including the lip, the gum, the inner aspect of the cheek, the tongue, and the floor of the mouth, is a rare site for benign tumours; but it is comparatively a common site for carcinoma.

Carcinoma of the tongue is a common lesion and accounts for more than a half of all intraoral carcinomas.

Predisposing factors

1. Chronic irritation – caused by:
 - Sharp tooth or an ill-fitting denture leading to repeated trauma.
 - Smoking – particularly by a clay pipe.
 - Spirits – excessive alcohol intake.
 - Spices.
 - Oral sepsis.

2. Syphilis – syphilis and carcinoma often coexist. Syphilis can cause leukoplakia which is a precancerous condition. *The carcinoma developing upon this leukoplakic patch is often fissured in type and radio-resistant, so it should always be treated by surgery.*
3. Chronic superficial glossitis, especially when it has reached the leukoplakia stage.
4. Sessile papilloma of the tongue.
5. Plummer–Vinson syndrome in case of females.

In short, *the predisposing factors can be remembered by six S's* – Sharp tooth, Smoking, Spirits and spices, Sepsis, Syphilis and Superficial glossitis.

Surgical pathology

Site of election (Fig. 8.3)

1. Anterior two-thirds of the tongue at or near the lateral margin – 25 percent $\times$ 2.
2. Posterior one-third of the tongue – 20 percent.
3. Tip of the tongue – 10 percent.
4. Dorsal surface of anterior two-thirds of the tongue – 10 percent.
5. Ventral surface of anterior two-thirds of the tongue – 10 percent.

The tip or the centre of the tongue is less commonly affected by carcinoma unless it arises in gummatous ulcer in the region.

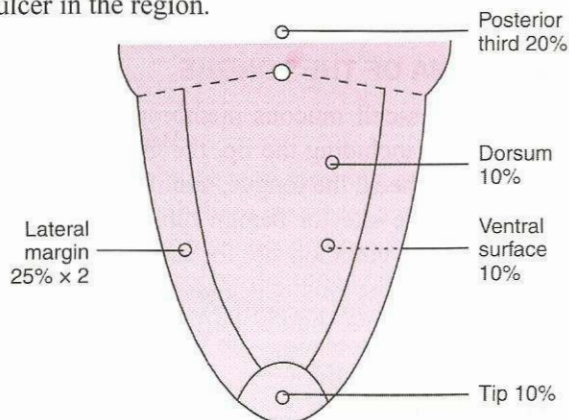


Fig. 8.3. Site of election for carcinoma of the tongue.

Macroscopic features

The growth may appear as:

1. *Ulcerative type* – with raised, irregular, rolled or everted margin, and a sloughing yellow grey base and induration of the surrounding tissues.

2. *Papilliferous or warty type* – varying from a small papilloma with broad and indurated base and covered with an excess of proliferating filiform epithelium to a large verrucous or cauliflower-like mass.
3. *Nodular type* – a submucous nodule or plaque.
4. *Fissured or cracked type with induration* – chronic in nature with no signs of healing, usually follows chronic superficial glossitis or syphilis.
5. *Frozen type* – here the tongue is transformed into an indurated mass with restricted mobility – ankyloglossia.

Microscopic features

Depend largely on which part of the tongue is affected:

1. *Anterior two-thirds of the tongue:* Squamous-cell carcinoma with characteristic cell-nest formation with the inner older cells showing keratinization and the outer younger cells arranged in an 'onion-peel-like' manner.
2. *Posterior one-third of the tongue:* Lympho-epithelioma or basal-cell carcinoma or transitional-cell carcinoma.
3. Rarely an adenocarcinoma may also occur.

Spread

1. Local spread

Carcinoma of the anterior two-thirds of the tongue, which starts usually on the lateral margin – invades the floor of the mouth early; but spread seldom extends across the midline.

Invasion of the floor of the mouth causes thickening of local tissues and reduces mobility of the tongue. Spread may extend to the mandible.

Carcinoma of the posterior one-third of the tongue – tends to spread to the corresponding tonsil, epiglottis, the soft palate, and even the vocal cord.

2. Lymphatic spread

Common; the lymph nodes are enlarged and very hard. The lymph nodes are involved by embolic spread (due to squeezing off of the malignant cells by the activity of the tongue musculature), not by permeation, so that the intervening tissue is not involved.

The lymphatics drain in the following way:

- *From the tip* – drain into the submental lymph nodes of both the sides.
- *From the anterior two-thirds* – drain unilaterally to the submandibular nodes and then to the lower deep cervical chain or juguloomohyoid nodes.
- *From the posterior one-third* – drain into the upper deep cervical chain or jugulodigastric nodes of both the sides.

Ultimately all the lymph drainage from the tongue reaches the jugulo-omohyoid lymph nodes in the deep cervical chain (Fig. 8.4A & B).

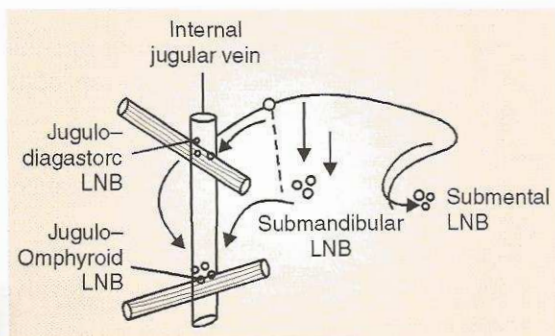


Fig. 8.4A. Lymph drainage of the tongue.

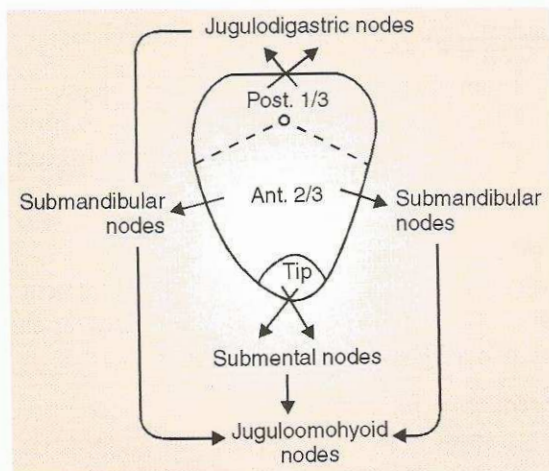


Fig. 8.4B. Lymph drainage of the tongue.

Points to note:

- It is important to note that in 50 percent of cases, the lymphatic vessels draining the anterior two-thirds of the tongue and the floor of the mouth traverse the periosteum of the mandible on their way to submental and submandibular lymph nodes.

- It is also to be remembered that the septic infection which invariably occurs in the malignant ulcer, may cause a non-malignant enlargement of the lymph nodes under the jaw. Such nodes are tender and firm and respond to antibiotics.
- Because of the secluded position and consequent late diagnosis, growths of the posterior third of the tongue show the highest incidence of cervical node metastases.

3. Blood spread

Very rare, occurs in only 2 percent of patients and in these cases the neoplasm is situated in the extreme posterior third of the tongue in almost every instance.

Diagnostic criteria

1. *Age*: Common in fifth or sixth decades.
2. *Sex*: Sex incidence is now equal.
3. *Symptoms*: In the early stage the patient complains of a *painless lump, irregularity, or ulcer on the surface of the tongue*.

Sadly, many a patient fails to notice or else disregards the lesion in its early stage and may report to the doctor only because of one or more of the following symptoms:

- Enlarging ulcer causing pain in the tongue; the pain is burning in nature.
- Pain referred to the ear (this is due to involvement of the lingual nerve and the pain is referred via the auriculotemporal nerve which is also a branch of the mandibular division of Trigeminal nerve).
- Excessive salivation: The saliva may be blood-stained. Excessive salivation is due both to excessive secretion from irritative lesion of the tongue and to difficulty in swallowing.
- Difficulty in swallowing and mastication.
- Foetor oris – due to bad oral hygiene and secondary bacterial stomatitis. The patient becomes offensive to his associates.
- Ankyloglossia and difficulty in speech – due to extensive carcinomatous infiltration of the tongue and/or floor of the mouth.
- Alteration of the quality of the voice – early with the lesions at the back of the tongue.
- Dysphagia – particularly in the lesions at the back of the tongue.
- General features of malignant cachexia, anaemia, anorexia, progressive loss of body weight – in late stages.

4. Alternatively, the patient may present with a lump in the neck (due to secondary deposits in the cervical lymph nodes) before he has noticed any abnormality in the tongue.
5. Previous history of any venereal disease, or any trouble with the teeth or dentures should be asked. Smoking and tobacco chewing habits should be enquired.



Fig. 8.5. Flat type of malignant growth on the left lateral margin of the tongue extending also on the undersurface of the tongue.



Fig. 8.6. Malignant ulcer on the left lateral margin of the tongue. (Courtesy: Prof. Sudeb Saha, MS, FAMS)

Examination:

6. Local examination: Note the site and character of the lesion on the tongue (see macroscopic features). Palpate for the *induration* of the lesion. Note the *mobility* of the lesion, and of the tongue.
7. It is also important to examine the posterior part of the tongue, floor of the mouth, tonsils and fauces, gums and teeth, and jaw.
8. Examine the neck for lymph node enlargement.

Note: Lesions in the posterior third of the tongue are difficult to diagnose in early stage. Usually presents when large in size causing difficulty in swallowing.

Change of voice may be the earliest symptom. Presence of multiple enlarged neck nodes (secondaries) is the commonest symptom. *Early indirect laryngoscopy is the best way of early diagnosis.*

Special investigations

1. Biopsy – must be performed before any treatment is planned.
Wedge biopsy from the edge of the ulcer can be taken under local anaesthesia. In case of proliferative growth punch biopsy is recommended. In case of posterior third growth, biopsy taken under general anaesthesia with the help of laryngoscope.
2. Laryngoscopy – to see the posterior one-third of the tongue, especially the region of the vallecula.
3. Blood for W.R. and Kahn – to be done in every case and, if positive, anti-syphilitic treatment should be started.
4. X-ray of the mandible preferably orthopantomogram in every case to detect any bony involvement, erosion.
5. Chest X-ray to detect any rare pulmonary metastasis.

Differential diagnosis

1. From other types of ulcers of the tongue.
2. From the rare tumours of the tongue – papilloma, haemangioma, lymphangioma, neurofibroma, lingual thyroid.

Treatment

Either surgery or radiotherapy is the treatment of choice. Results of surgery or radiotherapy for early carcinoma of tongue are equivalent.

Preliminary measures

1. A swab from the ulcer and the mouth is taken for culture and antibiogram to determine the predominant organism, and the most suitable antibiotics to be used.
2. Maintenance of oral hygiene by frequent antiseptic mouth washes.
3. Extraction of any sharp or carious tooth.
4. Suitable antibiotics (according to sensitivity test) to be given to control and prevent secondary infection.

Radical treatment

It includes:

1. Management of the primary lesion.
2. Management of the involved neck glands.

Management of the primary lesion (Fig. 8.7)

- I. *Growth in the anterior two-thirds of the tongue:* Wide excision is the preferred choice.

1. *Carcinoma in situ (CIS) or the lesion is ≤ 1 cm in size:* Wide excision of the growth with 1 cm clearance both in margin and depth is to be performed. Reconstruction of the tongue is not necessary.

The excised tissue should be sent for histopathological examination. If it shows either a CIS or an early invasive carcinoma which has of course been adequately excised – no further treatment is necessary but monthly follow-up is required upto 6 months.

2. *Growth is between 1 and 2 cm in size –* Treated by *partial glossectomy* with 2 cm clearance around the tumour. The defect may be grafted with split skin.

3. *Growth is between > 2 to 4 cm in size –* Options are:

- A. *Primary radiotherapy:* Usually external beam irradiation is preferred. Radiotherapy is successful in majority of cases depending on the site and type of the tumour. If radiotherapy fails to cure within 2 months of completion of treatment – surgery is indicated.

- B. *Surgery:*

- (i) *Hemiglossectomy:* Removal of around 50% of the tongue on the side of the tumour.

- (ii) *Sub-total glossectomy:* Removal of anterior two-thirds of the tongue when the tumour is central.

The defect may be grafted with split skin.

4. *When the growth is more than 4 cm or reaches or involves within 2 cm of the mandible:* Removal of that half of the mandible (hemi-mandibulectomy) is to be done. A stainless steel wire may be used to hold the remaining half of the jaw in position.

In these cases radiotherapy may not be successful. Moreover, radiotherapy causes necrosis of the mandible.

- II. *Any growth in the posterior third of the tongue, whether less or more than 1 cm in diameter, is treated by Teletherapy, because this secluded position is anatomically difficult both for surgery and for interstitial irradiation therapy.*

- III. *When the carcinoma of the tongue is associated with syphilis – they should be treated with surgery, because these cases are radioresistant.*

Management of the lymph nodes

Depends upon whether they are palpable or not. If palpable, whether mobile or fixed.

- I. *If the lymph nodes are impalpable or clinically normal* then prophylactic excision is unnecessary, but a careful regular monthly checkup is required.
- II. *If the lymph nodes are palpable but mobile*—it may be due to infection, with or without metastasis.

So, a course of antibiotics for 3 weeks is given postoperatively. If no response, block dissection of the neck is done preceded by FNAC of the nodes.

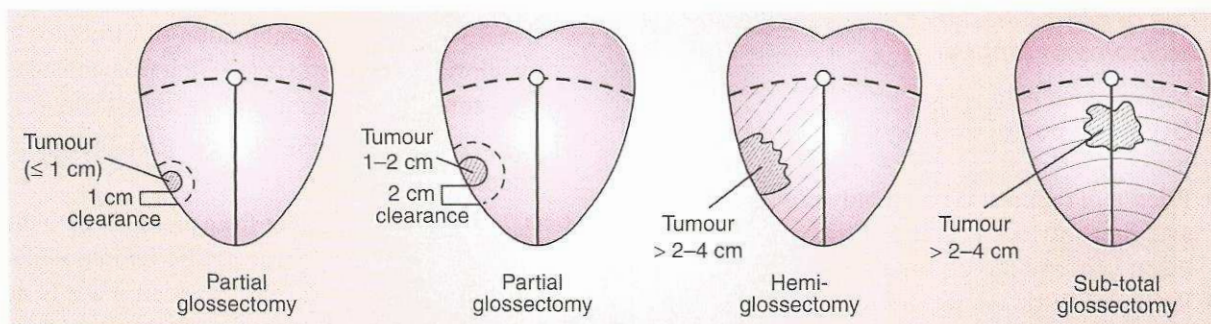


Fig. 8.7. Surgical management of primary tumour of the tongue.

III. *If the lymph nodes are palpable, firm to hard, mobile and they are suspected of being involved due to metastasis*—block dissection of the neck is done preceded by FNAC of the nodes to confirm diagnosis.

The block dissection of the neck may be combined with hemiglossectomy and hemimandibulectomy in the same sitting (**Commando operation**).

IV. *If the lymph nodes are fixed* then surgery is contraindicated and radiotherapy is used.

Radiotherapy

(a) *Interstitial irradiation (Brachytherapy)*: For small primary tumours – may be curative.

For large primary tumours – initial radiotherapy may be given to reduce the tumour size so that the resection will be better performed later.

Radium needles, radon seeds, radioactive tantalum wires or ^{192}Ir wires are used. The average dose in carcinoma of the tongue is between 6,000 r and 8,000 r.

The needles are inserted vertically in the substance of the tongue in one plane at 1 cm apart. The eyes of the radium needles are placed below the mucous membrane and are tied with one another by a silk thread. The free end of the thread is fixed to the face by leucoplast. The needles are kept in situ for 7 to 10 days.

(b) *Teletherapy* (external beam radiation with a telecobalt or linear accelerator): The form usually used is cobalt 60 unit. It is indicated particularly for the growth of the posterior third of the tongue and for recurrent growths after adequate excision as palliative mode.

Role of chemotherapy in carcinoma of tongue

- Not of much help.
- Given in postoperative period and also for palliation.
- Price-Hill regimen is commonly used. Drugs used are methotrexate, vincristine, adriamycin, bleomycin and mercaptopurine.
- Regional intra-arterial administration (through external carotid artery using arterial pump) of

Amethopterin in the dose of 50 mg per day for 5 days can be used, either as a preliminary to excision or as a palliative measure.

Palliative treatment in carcinoma of tongue

1. In case of large and fixed primary growth – radiotherapy will be tried.
2. In case of recurrence after radiotherapy or surgery – cryosurgery may be tried.
3. In case of extreme pain due to advanced growth – blocking of the trigeminal nerve with 5% phenol may be tried.

Prognosis

This depends on the site and stage of the growth, and lymph node involvements.

1. Site:
 - Growth in Anterior two-thirds – 50 percent 5 years survival rate.
 - Growth in posterior one-third – 10 percent 5 years survival rate.
2. Stage:
 - Early stage – 60 percent 5 years survival rate.
 - Late stage – 15 percent 5 years survival rate.
3. Nodes:
 - If not involved – 60 percent 5 years survival rate.
 - If involved – 15 percent 5 years survival rate.

Terminal events

If unsuccessfully treated, the disease turns inevitably to a fatal course. Death occurs usually in one of the following ways:

1. *Haemorrhage* from the primary growth and also from the carotid artery or internal jugular vein when eroded by metastatic lymph nodes.
2. *Combined malignant cachexia, starvation and exhaustion* from a combination of difficulty in swallowing and anorexia resulting from infected, foul-smelling fungating ulcer within the mouth.
3. *Aspiration bronchopneumonia*, from the super-added oral sepsis.
4. *Asphyxia* resulting either from pressure upon the upper air passages by metastatic lymph nodes, or from oedema of the glottis which is due to an extension of the lymphatic oedema around a growth at the back of the tongue.

Table 8.1. Common ulcers of the tongue and their differential diagnosis

	Dyspeptic	Traumatic	Tuberculous	Syphilitic (Tertiary)	Malignant
1. Age	Any age	Any age	Young adults	Young adults	Elderly
2. Site	Usually at the tip but may occur at any site with or without ulcers in the lip or cheek	Usually at the margin of the tongue, commonly towards the back	May occur at any site – margin, tip or dorsum of the tongue	Dorsum of the tongue mostly towards the base usually in the midline	Usually at the margin, common in anterior two-thirds of the tongue. May occur on the dorsum when superimposed on chronic superficial glossitis
3. Symptoms	Features of dyspepsia	Presence of a sharp tooth or ill-fitted denture causing repeated trauma	Features of tuberculous toxæmia	History of exposure, abortion or miscarriage	Excessive salivation, difficulty in articulation and speech, ankyloglossia, lump in the neck
4. Pain and tenderness	Exquisite	Marked	Exquisite	Painless	Early – painless. Late – painful and may be referred to ear
5. Shape	Small and circular	Any shape according to the shape of traumatic agent	Oval or circular	Oval or circular	Irregular
6. Number	Single or multiple	Single	Multiple	Single	Usually single; may be multiple when superimposed on chronic superficial glossitis
7. Depth and size	Shallow and small	Moderate	Shallow and moderate	Deep and may be large	Early—Shallow and small. Late—Moderate
8. Edge of the ulcer	Oedematous	Oedematous	Undermined	Punched out	<i>Rolled out and everted</i>
9. Floor	Covered by red granulation tissue	Covered with slough	Covered with pale granulation tissue	Covered with washed leather slough	Covered with necrotic debris and looks dirty grey
10. Discharge	Thin and watery	Often purulent	Thin and watery	Greyish white	Products of necrosis, thick and sometimes purulent. The discharge is offensive
11. Induration in the base and in the wall	Nil	Very slight	Nil	Very slight	Marked

(Contd.)

	Dyspeptic	Traumatic	Tuberculous	Syphilitic (Tertiary)	Malignant
12. Neck lymph nodes	Nil	Nil unless secondarily infected when the lymph nodes are firm and tender	Enlarged and matted with or without cold abscess	Enlarged, shotty and discrete. There may be enlarged epitrochlear and occipital lymph nodes	Early—Nil. Late—Enlarged and stony hard, may be fixed
13. Investigation	Stool	For presence of sharp tooth or ill-fitted denture	1. Routine blood for Hb% T.C., D.C. and fasting E.S.R. 2. Chest X-ray 3. Sputum for AFB 4. Mantoux test 5. Examination of the ulcer under magnifying glass—ulcer appears to be surrounded by minute sentinel tubercles	Blood for W.R. and Kahn	Blood for W.R. and Kahn
14. Response to treatment	Responds to treatment of dyspepsia	Usually heals after removal of source of irritation e.g., sharp tooth	Responds to anti-Koch's treatment	Responds to anti-syphilitic treatment	No response to conservative treatment. Surgery is the method of choice in most cases

Post-pertussis ulcer:

1. Occurs in children following whooping cough.
2. Ulcer is confined mostly on the frenum linguae.

Herpetic ulcer:

1. Common in children and young adults
2. Occurs due to herpetic affection of the lingual nerve.
3. To start with there is unilateral acute neuralgic pain on the affected side.
4. Followed a few hours later by appearance of a vesicle which later on bursts to form a very painful superficial ulcer.

Simple ulcers due to glossitis:

1. Ulcer is usually single, known as Smoker's patch.
2. Occurs in chronic superficial glossitis.
3. Burning pain during taking of food.

LIPS

Pigmented lips

Causes

1. Peutz-Jeghers' syndrome
2. Addison's disease

Macrochelia

Causes

1. Lymphangioma — commonest
2. Haemangioma
3. Chronic inflammation

Neoplasms

Benign

Usually uncommon and of following:

- Naevi
- Papilloma
- Fibroma
- Lipoma
- Haemangioma
- Lymphangioma
- Pyogenic granuloma
- Keratoacanthoma
- Leukoplakia
- Ectopic salivary tumour.

Malignant

1. Squamous cell carcinoma — commonest
 2. Basal cell carcinoma
 3. Melanoma
 4. Adenocarcinoma.
- } Rare

CARCINOMA OF THE LIPS

Squamous cell carcinoma is common in the lip, affecting almost exclusively the lower lip.

Incidence (Fig. 8.7)

Lower lip	93 percent
Upper lip	5 percent
Angle of the mouth	2 percent

Site of lesion

The carcinoma may arise at:

- the junction of the skin and mucous membrane of the lip;

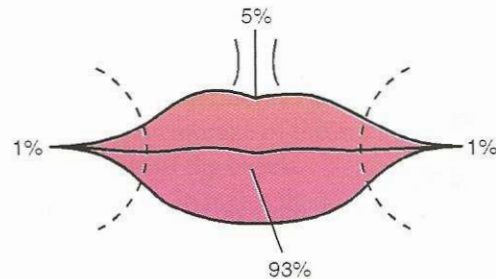


Fig. 8.8. Incidence of carcinoma lip.

- the mucous membrane of the 'true lip' i.e., the area between the red margin and the area of contact between the two lips;
- the mucous membrane covering the inner side of the lip – so during examination the lip should be everted to expose the inner surface.

Predisposing factors

1. Aged males between 60–70 years.
2. Usually an outdoor worker, who is continuously exposed to strong sunlight, wind-drift and driving rain.
3. Recurrent blistering cheilitis (Inflammation of the lips) – 40 percent cases.
4. Addiction to 'Khainy' – 'Khainy' is a mixture of raw tobacco and slaked lime. In many parts of India, people are in the habit of keeping Khainy between the lower lip and gum and thus make the lower lip especially vulnerable to carcinoma.

Origin

1. De novo
2. On some pre-cancerous conditions, e.g.,
 - Leukoplakia
 - Chronic fissures at the angle of the mouth
 - A sessile papilloma.

Macroscopic features

Carcinoma of the lip may present as:

- a nodule or fissure
- a typical malignant ulcer
- a warty papilliferous tumour
- an infiltrative induration.

Microscopic features

Most of the carcinomas are squamous cell carcinoma with 'cell-nests' and prickly cells.

Rarely basal-cell carcinoma, adenocarcinoma or malignant melanoma may occur.

Spread

Local spread

It may occur and it is more on the surface than in the substance of the lip. Because of close contiguity, the jaws may be invaded directly.

Lymphatic spread

- From the lower lip – to the submental and submandibular lymph nodes, and later to the jugulo-digastric lymph nodes.
- From the upper lip – spread occurs early to the pre-auricular, submandibular and later to the deep cervical lymph nodes.
- From the angle of the mouth – spread occurs both above and down, and so carcinoma situated at this site is more malignant.

Metastasis, however, is slow, and 80 to 85 percent patients show no sign of lymph node metastasis within the first year.

Blood spread

Very rare.

Special investigation

Blood for W.R. and Kahn – to exclude syphilis, because carcinoma may supervene on pre-existing syphilitic sore. *Carcinoma supervening on a syphilitic sore is almost always radioresistant.*

Differential diagnosis

1. Simple papilliferous wart – which may itself be a pre-malignant condition.
2. Keratoacanthoma.
3. Chancre: The lip is the commonest extra-genital site for a chancre. It is accompanied by local oedema and enlarged regional lymph nodes. Carcinoma may supervene on pre-existing syphilitic sore.
4. Haemangioma.
5. Lymphangioma.
6. Herpes simplex.
7. Mucous cyst.



Fig. 8.9. Carcinoma lip involving the right half of the lower lip extending to the angle of the mouth. (Courtesy: Prof. Diptendra Sarkar, MS, DNB, FRCS)



Fig. 8.10A. Carcinoma lip involving both the lower lip and upper lip. The upper lip lesion becomes more visible on everting the upper lip – "Kissing ulcer". No lymph node metastases were noted in the neck.



Fig. 8.10B. Same patient. The upper lip everted.

Treatment

It includes:

1. Management of the primary lesion
2. Management of the lymph nodes.

Management of the primary lesion

1. In early cases without glandular metastasis – *radiotherapy is the treatment of choice* and the prognosis is extremely favourable.
2. *Excisional surgery is indicated:*
 - If radiotherapy is not available.
 - When the patient is unable to attend the radiotherapy clinic regularly as he resides in a place which is far away from the clinic.
 - In late cases.
 - Recurrence after irradiation.
 - Carcinoma lip invading the lower jaw.
 - Carcinoma supervening on a leukoplakic patch.
 - Carcinoma supervening on syphilitic sore.

Principle: The tumour is removed together with a 1 cm margin of surrounding healthy tissue, by means of a 'V' shaped incision which includes the full thickness of the lip.

When a more radical excision is indicated some form of plastic surgery are adopted for construction of a new lip (swinging or rotation flaps from the cheek or neck).

Management of the lymph nodes

1. *If not palpable* – only observe at a regular interval of one month.
2. *If palpable, mobile, but not hard* – it may be due to infection. So, a course of antibiotics for 3 weeks is given. If no response – block dissection.
3. *If palpable, hard and mobile*, and suspected to be due to metastasis – block dissection.
4. *If palpable and fixed* – radiotherapy.

Prognosis

80 percent 5 years survival rate with surgery or radiotherapy when cervical lymph nodes are not involved.

CARCINOMA OF THE CHEEK

- Squamous cell carcinoma can occur in any part of the mucous membrane of the mouth including the inner aspects of the cheek.
- More common in males than in females.
- Commonly found in Eastern races who are in the habit of chewing the betel-nut and store the squid thereof in the cheek. Also common in people who use 'Khainy', roasted tobacco.
- In Western countries, it is more common among the people who smoke heavily and drink alcohol.
- The early symptoms are minimal and so are frequently ignored until there is pain and difficulty in speaking or eating. Then the patient may present with a *typical malignant ulcer, a fissure or a warty papilliferous growth* with or without foul-smelling discharge.
- The majority are slowly growing. The spread occurs by local infiltration which often transgresses nearby anatomical boundaries.
- Cervical lymph nodes are commonly involved following a secondary spread. Distant metastasis is rare and late, but local recurrence rate is high.

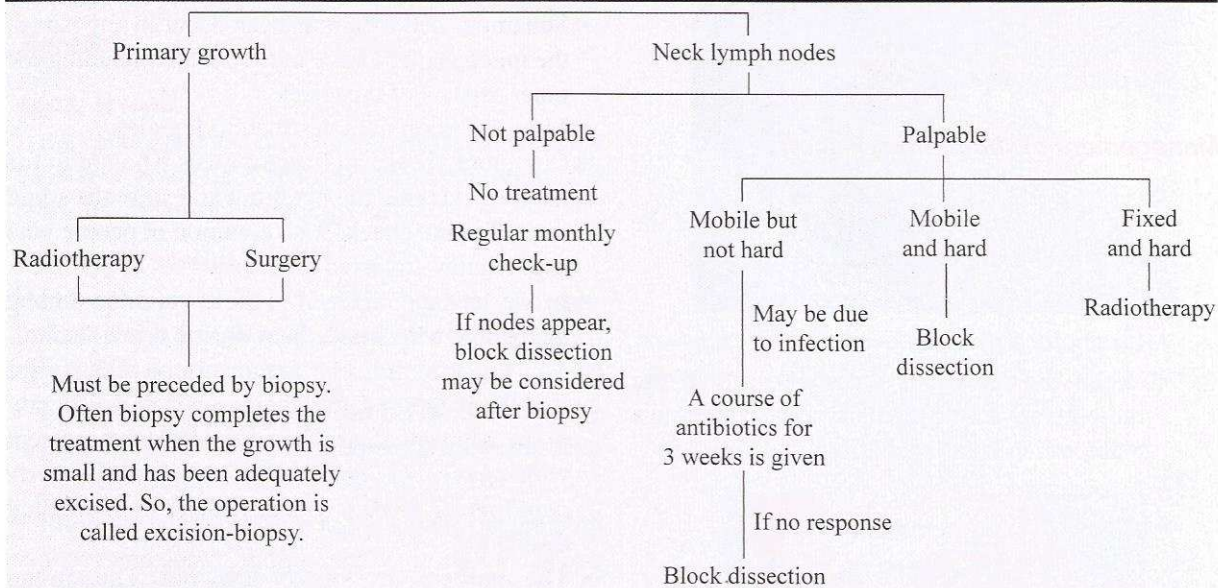
Treatment

1. *Radiotherapy:* Squamous cell carcinoma of cheek is radiosensitive. In early cases without lymph node metastasis – radiotherapy may be tried first after confirmation of biopsy. *Interstitial irradiation* may be given by Iridium wire. *External irradiation* is given by megavoltage machines.
2. *Surgery:* Excisional surgery is indicated in:
 - Meagre facilities of radiotherapy
 - Late cases
 - Recurrence after radiotherapy
 - Residual tumours.

The resulting defect following excisional surgery is made good by rotational flap, reflecting a flap of skin from the temporal region or by pedicle grafts. When the skin from the temporal region is taken the buccal aspect of the cheek becomes lined by the skin.

3. If cervical lymph nodes are involved – block dissection is required.

Outline of Treatment of Carcinoma Lip, Tongue and Cheek



9

CHAPTER



Umbilicus and Abdominal Wall

UMBILICUS

The umbilicus is represented by a depressed scar in the midline of the anterior abdominal wall. It is a deficiency formed due to meeting of the four folds of embryo – head-fold, tail-fold and two lateral folds. Through this deficiency, the umbilical cord connects the foetus with the mother. This deficiency transmits two sets of structures:

1. Vitelline components
2. Body stalk components.

Vitelline components consist of:

- the vitelline duct;
- the vitelline vessels;
- the splanchnic layer of the intra-embryonic mesoderm forming the coverings of the vitelline structures.

Body stalk components consist of:

- allantoic diverticulum;
- umbilical vessels;
- extra-embryonic mesoderm of the connecting stalk which gives rise to Wharton's jelly by mucoid degeneration.

VITELLOINTESTINAL DUCT

It is a tubular outgrowth of the yolk sac which connects the extra-embryonic part of the yolk sac with primitive mid gut. During the 12th week of the

intra-uterine life when the gut returns from the extra-embryonic coelom the vitellointestinal duct separates from the intestinal loop and ultimately disappears.

Anomalies

The vitellointestinal duct occasionally persists and gives rise to following complications (Fig. 9.1).

1. It remains patent throughout its whole length giving rise to umbilical fistula – intestinal fistula (Fig. 9.1A).

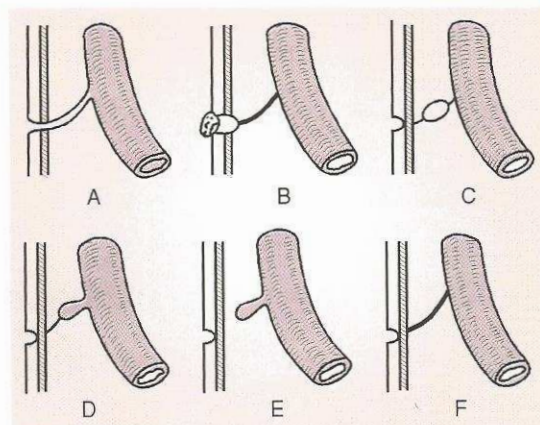


Fig. 9.1. Different anomalies of vitello-intestinal component of umbilicus. A, intestinal fistula; B, umbilical adenoma; C, enterocystoma; D & E, Meckel's diverticulum; F, remnant of V.I. duct persists as intraperitoneal band.

- The fistula discharges mucous, and rarely faeces – faecal fistula. The discharge is usually noticed in the first few weeks or months of life.
 - Occasionally, intestinal parasites such as *Ascaris lumbricoides* may extrude from the umbilicus.
 - It may be possible to cannulate the fistula. The catheter goes into the intestines.
 - There could be double intussusception through the umbilicus rarely.
 - The fistula may close spontaneously when the lumen of the communicating tract is narrow.
 - Sometimes the fistula persists, when the abdomen should be explored, and the patent duct should be excised in toto along with a segment of ileum.
2. Sometimes small portions of the duct near the umbilicus remain patent, the rest being obliterated. This gives rise to a sinus that discharges mucous. The epithelial lining of this patent sinus may be everted to form an *adenoma* or *raspberry tumour* (Fig. 9.1B).
 3. Sometimes both the umbilical and the intestinal ends of the duct become obliterated, but the central portion remains patent and gives rise to an intra-abdominal cyst – *enterocystoma* (Fig. 9.1C).
 4. Sometimes the patency of the intestinal end of the vitellointestinal duct gives rise to *Meckel's diverticulum* which may or may not be attached to the umbilicus (Fig. 9.1 D & E).
 5. With its lumen obliterated or unobliterated the vitellointestinal duct provides an *intraperitoneal band* which is a potential danger, for intestinal obstruction is liable to occur. The obstruction results from a coil of small intestine passing under or over, or becoming twisted around, the band (Fig. 9.1F).
 6. Such a band may contract and pull a Meckel's diverticulum into a congenital umbilical hernia.
 7. Rarely the vitellointestinal remnant connected to the Meckel's diverticulum, but not attached to the umbilicus, become adherent to, or knotted around another loop of small intestine, and so causes intestinal obstruction.
 8. Very rarely a cord like structure may stretch from the umbilicus across the ileum to end in the mesentery. This is an obliterated vitelline artery but not the vitellointestinal duct.

ALLANTOIS

It is a tubular diverticulum from the yolk sac passing from the cloaca through the umbilicus and the umbilical cord to end blindly at the placenta. *The part within the body forms the urachus and the bladder except the trigone.* The remainder disappears.

As the bladder descends into the pelvis the urachus becomes obliterated and turns into a fibrous remnant which is known as median umbilical ligament. This ligament lies between the transversalis fascia and the peritoneum.

Anomalies

1. The urachus occasionally remains patent and communicates with the umbilicus leading to umbilical fistula through which the urine is discharged – *urinary fistula*. Though the patent urachus is a congenital lesion, urinary fistula develops only when there is obstruction to the flow of urine in the distal urinary passage, i.e., the neck of the bladder to downwards, e.g., posterior urethral valve. A fistulogram will confirm the diagnosis. *The treatment is directed to remove the distal obstruction first*, which helps in the spontaneous closure of the fistula. If the leak still continues, umbilectomy and excision of the urachus down to the apex of the bladder are performed.
2. Rarely, the central portion of the urachus remains patent with the umbilical and vesical ends obliterated. This is a *urachal cyst*.

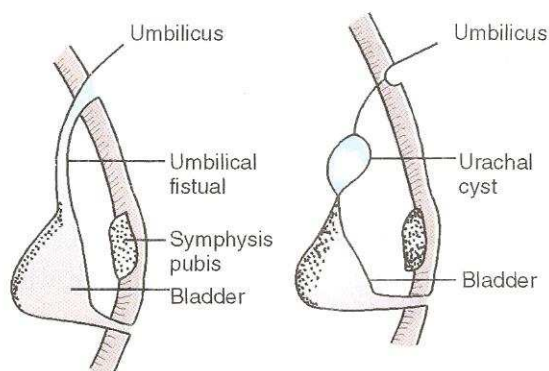


Fig. 9.2. Various anomalies of the allantois.

DISEASES OF THE UMBILICUS

1. *Umbilical hernia* – commonest.
 - Congenital
 - Exomphalos
 - Congenital umbilical hernia
 - Acquired
 - Infantile umbilical hernia
 - Adult umbilical hernia (See Chapter 10)
2. *Anomalies of vitelline components*
3. *Anomalies of body stalk components*
4. *Umbilical fistula*
5. *Umbilical sinus* – pilonidal sinus
6. *Inflammation*
 - Umbilical granuloma
 - Umbilical dermatitis or omphalitis
7. *Neoplasm*
 - Benign
 - Umbilical adenoma
 - Endometrioma
 - Malignant
 - Primary carcinoma
 - Secondary carcinoma
8. *Umbilical calculus*
9. *Eversion* (e.g., in ascites)

ENTEROCYSTOMA

It is a pathological condition arising from the patent central portion of the vitellointestinal duct (Fig. 9.1C). It gives rise to intra-abdominal cyst. Through a band it is fixed to the umbilicus and to the small intestine.

The patient presents with a small, spherical, mobile swelling deep to the umbilicus.

Treatment

Excision of the cyst along with the band to avoid future intestinal obstruction.

URACHAL CYST

It is a rare pathological condition arising from the patent central portion of the urachus with the umbilical and vesical ends obliterated. It also gives rise to intra-abdominal swelling.

The patient presents with a small immobile swelling in the hypogastrium deep to the umbilicus.

Treatment

Excision of the cyst.

UMBILICAL FISTULAE

Causes

1. *Faecal*:
 - Patent vitellointestinal duct – already discussed.
 - Neoplastic ulceration due to direct infiltration by carcinoma of the transverse colon.
 - Neoplastic ulceration infiltrating the underlying bowel.
 - Tuberculous peritonitis – which may give rise to serous discharge.
 - Crohn's disease.
 - Actinomycosis.
2. *Urinary*: Patent urachus – already discussed.
3. *Biliary*: Gallbladder perforating at the fundus and sometimes discharging stone also.
4. *Iatrogenic*: If a gauze piece was accidentally left behind in the abdomen.

UMBILICAL GRANULOMA

It is a chronic infection of the umbilical cicatrix following severance and tying off of the umbilical cord. By the end of first week after birth, the dry umbilical cord separates following tying off. Either delay in separation or inadequate desiccation of the cord remnant can permit infection at the base and lead to granuloma formation. This granuloma prevents the raw area from becoming epithelialised.

The baby presents with a pouting umbilicus surmounted by a red or pink, moist mass of granulation tissue. The granuloma discharges serous fluid. It is friable and bleeds on touch.

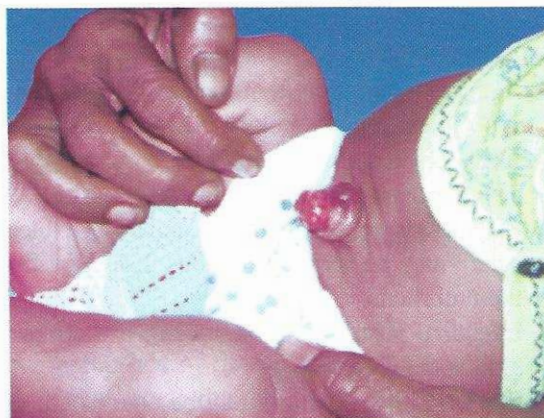


Fig. 9.3. Umbilical granuloma.

Differential diagnosis

Umbilical adenoma.

Treatment

Cauterisation: Either chemical cauterisation with silver nitrate stick or diathermy cauterisation followed by dry dressing usually destroys the umbilical granuloma. Excision is rarely necessary.

If the granuloma persists or recurs in spite of the above treatment, one should suspect an umbilical adenoma.

RASPBERRY TUMOUR

Syn.: Umbilical adenoma; Enteroteratoma

It is a pathological condition arising from the patent distal segment (i.e., near the umbilicus) of the vitello-intestinal duct (Fig. 9.1B). The mucosa of the patent duct prolapses through the umbilicus and gives rise to a soft pink tumour which looks like a raspberry, hence the name. Histological picture – usually consists of small bowel mucosa, i.e., columnar epithelium rich in goblet cells.

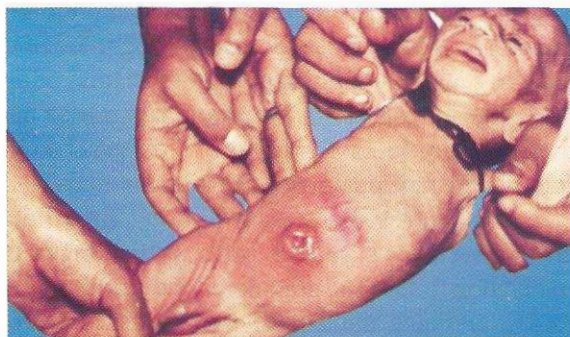


Fig. 9.4. Raspberry tumour.

Diagnostic criteria

1. *Age:* Infancy, rarely it may occur in later life.
2. A soft fleshy mass, pinkish in colour (raspberry-like), situated at the umbilicus.
3. The mass may be pedunculated or sessile.
4. The mass is moist with mucus and tends to bleed.
5. There may be serosanguineous discharge from the umbilicus.
6. Intestinal obstruction may occur, if there is an associated band.
7. Meckel's diverticulum may be co-existent and should be excluded by a barium meal, if possible.

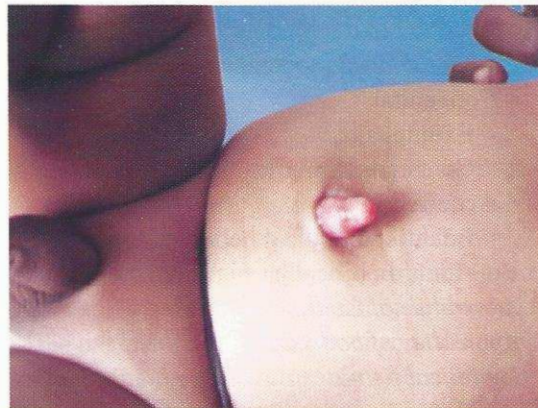


Fig. 9.5. Raspberry tumour – note the excessive prolapse of the mucosa from the patent distal segment.

Differential diagnosis

Umbilical granuloma.

Treatment

1. If the tumour is pedunculated – a ligature is tied around its base. In a few days the polypus drops off.
If there is recurrence – umbilectomy is indicated.
2. Some groups advocate umbilectomy as the first choice of treatment.
3. Others advocate exploration of the abdomen routinely, because the tumour sometimes is associated with a patent vitellointestinal duct or a vitellointestinal band along with the Meckel's diverticulum.

So, after exploration of the abdomen, the Meckel's diverticulum and the attached band or duct are excised and at the same time umbilectomy is done.

ENDOMETRIOMA OF THE UMBILICUS

It is a pathological condition where there is presence of ectopic endometrial glands in the umbilicus.

Diagnostic features

1. It occurs in women between 20 and 45 years of age.
2. The umbilicus becomes painful and *bleeds at each menstruation*.
3. There is a small fleshy mass at the umbilicus.
4. Occasionally, an umbilical endometrioma is associated with endometriomas in the uterus or ovary.

Treatment

When solitary, umbilectomy will cure the condition.

Causes of discharge from umbilicus

Nature of the discharge	Cause
Serous/purulent discharge	Umbilical granuloma Umbilical adenoma
Faeculent discharge (usually with faeculent smell)	Patent vitellointestinal duct
Clear fluid (smells of urine)	Patent urachus
Blood discharge	Raspberry tumour Endometrioma of the umbilicus Umbilical granuloma Umbilical calculus Secondary carcinoma of the umbilicus



Fig. 9.6. Umbilical fistula discharging watery discharge – ? urachal fistula. This patient gave history of good urinary outflow.

UMBILICAL CALCULUS

It is inspissated desquamated epithelium which collects in a deep recess of the umbilicus and gives rise to inflammation and often blood-stained discharge. The calculus is black in colour. Treatment is to dilate the orifice and extract the calculus. If recurrence – umbilectomy.

NEOPLASMS OF THE ABDOMINAL WALL

- Benign
- Malignant

Benign tumours of the abdominal wall may arise

from any of the anatomical structures contained in it. Examples are lipoma, fibroma, neurofibroma, haemangioma etc. These are treated in the same manner as in other locations.

The most characteristic benign tumour of abdominal wall is the Desmoid tumour.

Primary malignant tumours of the abdominal wall are extremely uncommon. However, rarely one may come across fibrosarcoma of the abdominal wall.

DESMOID TUMOUR

Greek: *desmos* = tendon; *eidos* = appearance

The tumour is so called because it looks like a tendon in its appearance and texture.

It is a benign fibrous tumour *arising in the musculo-aponeurotic structures of the abdominal wall, especially below the level of umbilicus (particularly in the sheath of the rectus abdominis muscle)*. It is notorious for recurrence in spite of apparently adequate excision. So, it is also known as '*recurrent fibroid of Paget*'.

The tumour may also be extra-abdominal when it can involve the musculoaponeurotic structures in the region of the shoulder, the medial side of the thigh, buttocks and chest.

Aetiology

Exact aetiology is not known.

1. Common in women (80%): Particularly middle aged multiparous women, the reason being repeated trauma caused by stretching of the muscle fibres during pregnancy.
2. May occur in the scars of old hernial or other abdominal operation scar.
3. A small haematoma of the abdominal wall due to trauma may be a precipitating factor. However, haemosiderin pigment is absent in this lesion.
4. In certain cases of familial polyposis coli this condition may be noted (Gardner's syndrome).

Pathology

1. It is essentially a benign, hard fibroma. It is composed of fibrous tissue and multinucleated plasmodial masses resembling foreign body giant cells.

2. It may be circumscribed – yet uncapsulated or diffusely infiltrating fibroma.
3. It may be so hard that it cracks when it is cut.
4. It is a slowly growing tumour, but it infiltrates the surrounding muscle. Rarely it may extend through the muscles of the anterior abdominal wall to involve the bowel or bladder.
5. There may be myxomatous change when the tumour increases in size rapidly.
6. *Recurrence is very common* even after wide excision – particularly high after excision of the extra-abdominal desmoids.
7. But there is *no tendency to metastasize*.
8. Very rarely may undergo low grade sarcomatous change.

Diagnostic criteria

1. Subject: Common in middle-aged women.
2. Presents with lower abdominal lump, commonly in relation to rectus sheath.
3. The lump is present on one side of abdomen and *does not cross the midline*.
4. The lump may be well circumscribed or diffusely infiltrating.
5. Parietal swelling as confirmed by rising test when the tumour becomes more prominent.
6. The mass is firm to hard, smooth, discrete and unattached to the skin.
7. Clinically it may be difficult to differentiate a desmoid tumour from a fibrosarcoma.
8. Diagnosis is confirmed by excision biopsy.

Treatment

1. Wide excision including a margin of surrounding

healthy tissue of at least 2.5 cm. The resulting defect is made good by using tantalum gauze or nylon or prolene mesh in the repair.

2. Radiotherapy is of controversial value.
3. Drug therapy:
 - Theophylline and chlorthiazide may be given as an adjunct to surgery to promote tumour shrinkage.
 - Indomethacin, a drug which inhibits prostaglandin synthesis, together with ascorbic acid may be given. It retards the tumour growth.

SECONDARY CARCINOMA OF THE UMBILICUS

Secondary carcinoma at the umbilicus is not very uncommon but its presence indicates an advanced, widespread malignant disease. The primary tumour is commonly in the abdomen, e.g., in the stomach, colon or ovary. Occasionally, a metastasis from the breast is located here.

The tumour cells reach the umbilicus by trans-peritoneal spread or via the lymphatics that run in the edge of the falciform ligament of the liver. Umbilical metastasis is usually associated with multiple peritoneal metastases.

Umbilical metastasis is present as a *nodule* at the umbilicus, in a patient who is losing weight and feels sick. *The metastatic nodule may ulcerate, bleed and become infected*. It may become attached to and infiltrate the underlying bowel. Necrosis of the tumour then can cause an acquired umbilical fistula – intestinal fistula discharging faecal matter.

10

CHAPTER

Hernia



HERNIA

A hernia is an abnormal protrusion of the whole or a part of a viscus through an opening in the wall of the cavity in which it is contained.

An *internal hernia*, also known as concealed hernia, is due to the protrusion of the viscus commonly intestine through an internal defect or into a peritoneal recess within the abdomen e.g.,

- Hiatus hernia
- Diaphragmatic hernia
- Hernia through the foramen of Winslow
- Hernia through the peritoneal recess around:
 - (a) Duodenojejunal junctions:
 - (i) Superior
 - (ii) Inferior
 - (iii) Lateral
 - (iv) Medial
 - (b) Caecum: Paracaecal hernia.
 - (i) Superior ileo-caecal
 - (ii) Inferior ileo-caecal
 - (iii) Retrocaecal.
- Hernia through a defect in the mesentery following bowel resection.

An *external abdominal hernia* is protrusion of a viscus from the peritoneal cavity through a weak part of the abdominal wall, either congenital or acquired.

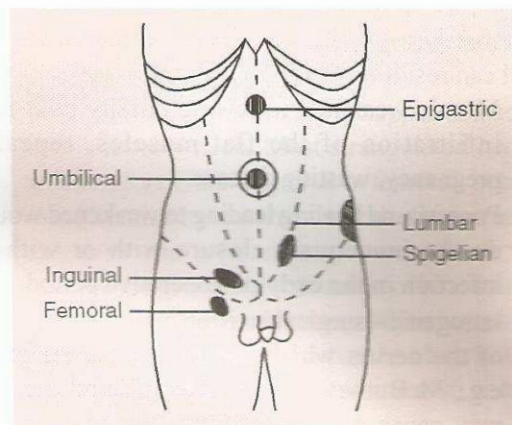


Fig. 10.1. External abdominal hernia.

Common external herniae are:

1. Inguinal
2. Incisional
3. Umbilical
4. Femoral

Less common types are:

1. Epigastric
2. Para-umbilical
3. Lumbar.

Rare types are:

1. Spigelian
2. Obturator
3. Gluteal.

Precipitating factors

Two factors are responsible for the production of a hernia:

- A. Weakness of the abdominal wall, and
- B. Continued or repeated increases in abdominal pressure.

A. Weakness of the abdominal wall

It may be congenital or acquired.

Congenital weakness: Due to persistence of congenital peritoneal sac, e.g.,

1. Persistence of processus vaginalis may lead to indirect inguinal hernia in males.
2. Patent canal of Nuck may lead to indirect inguinal hernia in females.
3. Incomplete obliteration of umbilicus may lead to umbilical hernia.

Acquired weakness: Weakness of the abdominal wall can result from:

1. Muscle weakness following obesity with fatty infiltration of the flat muscles, repeated pregnancy, wasting disease.
2. Poor wound healing leading to weakened wound due to inadequate closure with or without infection in the early postoperative period.
3. Iatrogenic – surgical incision may cause division of the nerves which causes muscle weakness e.g., McBurney's incision for appendicectomy may cause divisions of the subcostal or ilio-inguinal nerve leading to right sided direct inguinal hernia.

B. The increased intra-abdominal pressure

It becomes continued or repeated in following conditions:

1. Chronic cough, e.g., bronchitis.
2. Constipation.
3. Bladder neck or urethral obstruction, e.g. enlarged prostate or stricture urethra.
4. Severe muscular effort.
5. Parturition.
6. Vomiting.
7. Ascites.

Surgical pathology

An external abdominal hernia consists of:

1. A peritoneal sac

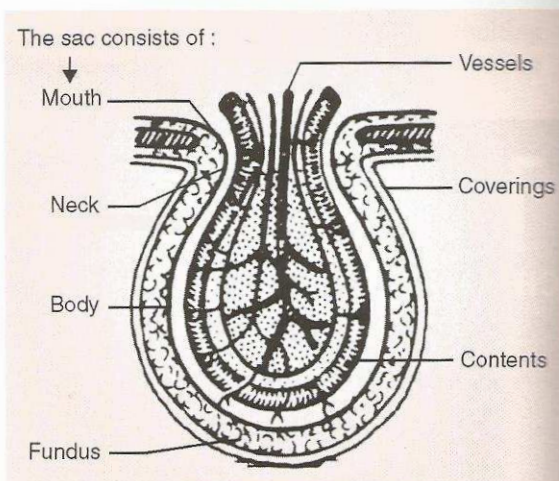


Fig. 10.2. Different parts of the hernial sac.

2. The coverings of the sac
3. The contents of the sac.

The sac is a pouch of stretched peritoneum which comes out through a gap in the abdominal musculature. The sac consists of four parts:

1. *The mouth* i.e., the opening of the sac through which the contents enter the sac.
2. *The neck of the sac* which is the most constricted part and passes through the abdominal musculature. Neck is narrow in indirect sac but wide in direct sac.
3. *The body* i.e., the main part of the sac holding the contents.
4. *The fundus* which is the most redundant and dependent part of the sac.

At first the sac is thin and delicate, but in long-standing cases the sac becomes considerably thick. Body of the sac is thin in infants and children.

The *coverings* are the layers of the abdominal wall which cover the hernial sac. These depend on the site and type of hernia and usually include skin and muscles of the abdomen.

The *contents* may comprise one of the following:

1. Omentum (omentoceles/epiplocele)
2. A loop of intestine (enterocele)
3. Meckel's diverticulum (Littre's hernia)
4. A part of the circumference of intestinal wall (Richter's hernia)
5. Two loops of intestine in the manner of 'W' (Maydl's hernia)

6. A part of bladder wall (direct/femoral hernia)
7. Ovary with or without fallopian tube
8. Fluid – slight amount of fluid almost always present but it increases in presence of ascites. The fluid may be blood stained when the hernia is strangulated.

Basic features of all herniae

1. They occur *through a weak spot* of the abdominal wall.
2. They have an *expansile impulse on coughing* (both visible and palpable).
3. They are *reducible* – either on lying down or with manual pressure. A reducible hernia is one in which the contained viscus can be returned to its original cavity.
4. A reducible hernia expands on coughing or straining – *cough impulse positive*.
In irreducible hernia – cough impulse is absent.
5. The contents of the hernia – commonly gut or omentum. In presence of gut, the hernia will feel soft and it may gurgle during reduction. In presence of omentum, the hernia feels doughy.

Complications of a hernia

1. *Irreducibility*: When the contents of the sac cannot be completely emptied from the sac because of:
 - adhesion formed between the contents and the sac or between the contents themselves;
 - growth of the omentum within the sac;
 - narrowing of the neck of the sac because of fibrosis, e.g., following continuous pressure of truss;
 - retention of faeces in the large intestine occupying the sac.

In a *simple irreducible hernia*, though the contents cannot be reduced, the blood supply remains intact, and there are no symptoms of intestinal obstruction. So the operation is not very urgent.
2. *Obstruction*: Irreducibility + obstruction of the lumen of the contained bowel leading to intestinal obstruction. The features are:
 - The hernia is *irreducible* but *painless*.
 - The sac is *lax and not tender*.

- *Cough impulse may be present.*
- *Features of intestinal obstruction are present.*
- Can precipitate strangulation if not treated early.

Hernia is one of the commonest causes of intestinal obstruction. So, hernial sites should always be examined in a patient presenting with intestinal obstruction.

3. *Strangulation*: Irreducibility + features of intestinal obstruction + arrest of blood supply to the contained intestine leading to gangrene. The features are:
 - The hernia is *irreducible* and *painful*.
 - The sac is *tense and tender*.
 - *Cough impulse absent*.
 - *Features of intestinal obstruction present* – pain abdomen, vomiting, abdominal distension, rebound tenderness.

Strangulation is more likely to happen in a hernia with a narrow neck. Most strangulated herniae are therefore either inguinal or femoral, because these herniae have narrow necks. *Often, the sac contains the greater omentum* which may become gangrenous due to arrest of blood supply showing the features of strangulation but without the features of intestinal obstruction. Strangulation without obstruction also noted in Richter's hernia.

4. *Reduction-en-masse*: Sometimes, during forceful manual reduction of irreducible hernia, the contents together with the covering sac gets pushed forcibly back into the abdominal cavity; the bowel within the sac may be strangulated by the neck of the sac. Thus the symptoms of obstruction or strangulation may not be relieved.
5. *Incarceration*. Incarcerated hernia is a *variety of irreducible hernia* where the content of the sac is large gut containing faeces. The large gut is fixed in the sac because of its size or adhesions. Here, the hernia can be indented like putty with the finger tip pressure because of the scybalous content of the gut. Though the hernia is irreducible, obstruction or strangulation rarely occur.
6. *Inflammation*, e.g., tuberculosis of the sac.

7. *Compression or torsion of the omentum within the sac.* The omentum may become gangrenous due to arrest of blood supply showing the features of strangulation but without features of intestinal obstruction.
8. *Collection of fluid in the hernial sac*—hydrocele of the hernial sac.
9. Maydl's hernia.
10. Sliding hernia.
11. Recurrent hernia.

INGUINAL HERNIA

This is the most common type of abdominal hernia in both males and females. There are two types:

1. Indirect or oblique inguinal hernia.
2. Direct inguinal hernia.

Surgical anatomy

Inguinal canal

The inguinal canal is an oblique fibromuscular tunnel about 4 cm long in the lower part of the anterior abdominal wall, a little above and parallel to the inner half of the inguinal ligament. It passes downward and medially and extends from the deep inguinal ring

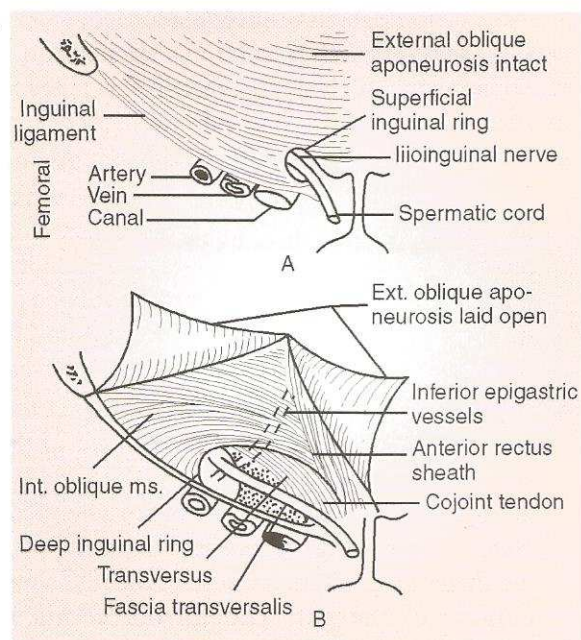


Fig. 10.3. Anatomy of the inguinal canal. A, external oblique aponeurosis intact; B, external oblique aponeurosis laid open.

to the superficial inguinal ring. The canal transmits the spermatic cord in males and the round ligament of uterus in females.

Boundaries

Anterior wall

1. Skin
2. Superficial fascia
3. External oblique aponeurosis – in its whole length
4. Internal oblique muscle – in its lateral third.

Posterior wall

1. Fascia transversalis – in its whole length
2. Conjoint tendon – in its medial half
3. Reflected part of inguinal ligament – in its medial third.

Roof

The arched fibres of the internal oblique and transversus abdominis muscles.

Floor

1. Upper grooved surface of the inguinal ligament.
2. Upper surface of the lacunar ligament – at the medial end.

Superficial inguinal ring

It is a slit in the external oblique aponeurosis situated 1 cm above and lateral to the pubic tubercle. It transmits the spermatic cord or round ligament, the ilio-inguinal nerve, the genital branch of the genito-femoral nerve and the processus vaginalis when it persists. The external oblique aponeurosis extends over the spermatic cord as external spermatic fascia.

Deep inguinal ring

It is a U-shaped condensation of the fascia transversalis being incomplete above. It is situated 1 cm above the midpoint between the anterior superior iliac spine and symphysis pubis. *Clinically the ring lies 1 cm above the point where the femoral artery passes under the inguinal ligament, i.e., 1 cm above the femoral pulse felt.* It transmits the spermatic cord or the round ligament, and the processus vaginalis when it persists. The fascia transversalis extends into the spermatic cord as internal spermatic fascia. The inferior epigastric artery courses up along the medial side of the ring.

The deep ring is the site through which indirect inguinal hernia occurs.

Hesselbach's triangle

This triangle is situated in the posterior wall of the inguinal canal. The triangle is bounded:

1. *medially* by the outer border of rectus abdominis muscle;
2. *laterally* by the inferior epigastric vessels;
3. *below* by the inner half of the inguinal ligament;
4. *floor* is formed by the fascia transversalis.

The triangle is almost bisected by the lateral umbilical ligament which is obliterated hypogastric artery. *Hesselbach's triangle is the site through which direct inguinal hernia occurs.*

Contents

1. In male: The spermatic cord.
2. In female: The round ligament of the uterus.
3. Ilio-inguinal nerve in both sexes.

Natural mechanisms preventing hernia

1. Obliquity of the canal.
2. Internal oblique muscle opposite the deep ring.
3. Shutter action of the arched fibres of internal oblique and transversus abdominis.
4. Plugging action of the spermatic cord due to contraction of the cremasteric muscle.
5. Sliding valve action of the U-shaped internal ring.

INDIRECT INGUINAL HERNIA

The hernia sac enters the inguinal canal through the deep inguinal ring lateral to the inferior epigastric artery. *The neck of the sac is lateral to the inferior epigastric artery.*

Indirect hernia is *usually congenital* due to persistence of patent processus vaginalis; it appears at or soon after birth or may appear in adolescence.

Indirect hernia *may be acquired* when the sac is formed as an outpushing of the abdominal peritoneum through the deep ring. The acquired variety may occur at any age in adult life due to increased intra-abdominal pressure following strenuous exercises, heavy weight lifting, straining at defaecation (chronic constipation) or at micturition

(enlarged prostate, stricture urethra) or at coughing (chronic bronchitis).

The indirect hernia may be subdivided into:

- Bubonocoele
 - Funicular
 - Vaginal or
- } Incomplete hernia
} Complete hernia

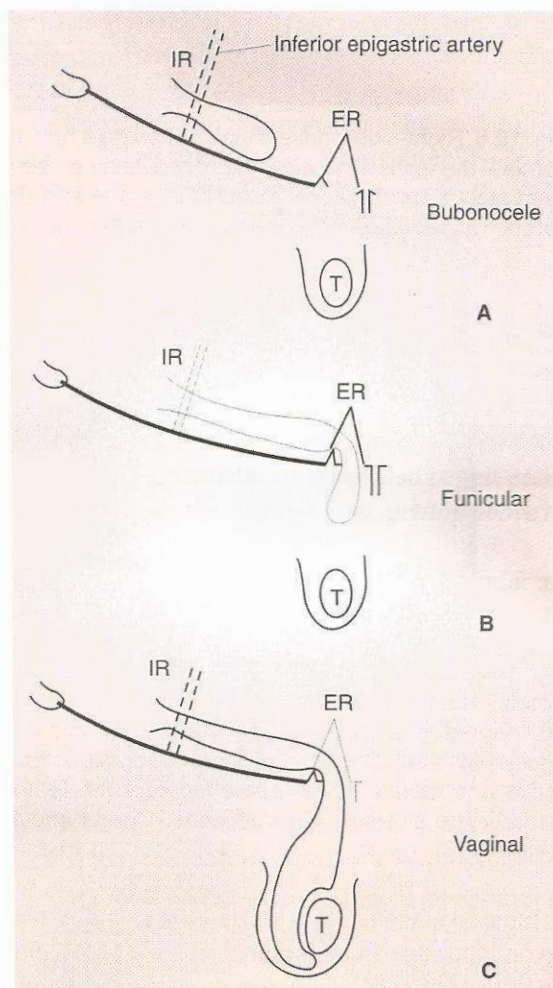


Fig. 10.4. Different types of indirect inguinal hernia.

Bubonocoele (Fig. 10.4A)

The processus vaginalis is closed proximal to the superficial inguinal ring. The hernia is limited in the inguinal canal and so *appears as an inguinal swelling.*

Funicular (Fig. 10.4B)

Here the processus vaginalis is closed at its lower end, so the sac of the hernia is separate from the sac



Fig. 10.5. Right-sided indirect inguinal hernia – funicular hernia – the testis lies below the hernia and can be felt separately from the contents of the hernial sac. This hernia was reducible. Cough impulse positive.



Fig. 10.6. Left incomplete indirect inguinal hernia in female. Note the site of the swelling—this is in the inguinal region above the groin crease i.e. above the inguinal ligament. On palpation the swelling will be found above and medial to the pubic tubercle (cf. femoral hernia). The swelling was reducible. Cough impulse positive.

of tunica vaginalis. The sac is closed and extends beyond the superficial inguinal ring but stops above the testis. *The testis lies below the hernia and can be felt separately from the contents of the hernial sac.*

Most of the indirect inguinal herniae belong to this category and are commonly seen in adults. It is usually acquired but may be congenital.

Vaginal hernia (Fig. 10.4C)

Also known as complete hernia. Here the processus vaginalis is patent throughout. Hence the sac is continuous with the tunica vaginalis of the testis. The hernia descends down to the bottom of the scrotum covering the testis in front and at the sides, *so the*

testis cannot be felt separately before reduction of hernial contents.

Both the funicular and vaginal herniae appear as inguino-scrotal swellings.

Indirect inguinal hernia is the most common hernia in children, adult and women.

Coverings of indirect inguinal hernia

From superficial to deep:

1. Skin
2. Superficial fascia. When the hernia comes out through superficial ring and descends into the scrotum – dartos muscle of the scrotum.
3. External oblique aponeurosis, or external spermatic fascia when the hernia comes out through the superficial inguinal ring.
4. Cremaster muscle.
5. Internal spermatic fascia.
6. Processus vaginalis or peritoneum – the sac.

DIRECT INGUINAL HERNIA

The hernia protrudes directly through the posterior wall of the inguinal canal in the Hesselbach's triangle. The direct hernial sac lies outside the cord (cf. indirect inguinal hernia) – behind, above or below the cord. *The neck of the sac lies medial to the inferior epigastric artery* – a differentiating point at operation from indirect hernial sac where the neck of the sac lies lateral to the artery.

Direct herniae are mostly acquired in nature. It is found predominantly in the elderly males due to increased intra-abdominal pressure leading to increased wear and tear of the conjoined tendon.

A direct inguinal hernia seldom comes out through the superficial inguinal ring and thus it is unusual for the sac to descend into the scrotum.

The neck of the direct hernial sac is wide and therefore the hernia appears immediately on standing and disappears again immediately when the patient lies down. *Moreover, because of this wide neck obstruction or strangulation is extremely rare.*

Coverings of direct inguinal hernia

From superficial to deep:

1. Skin
2. Superficial fascia
3. External oblique aponeurosis

4. Conjoint tendon when the sac passes medial to the lateral umbilical ligament
5. Fascia transversalis
6. Peritoneum – the sac.

Diagnostic criteria of an uncomplicated inguinal hernia

1. Age: It occurs in all ages, from birth to elderly. Indirect hernia is more common in younger and adult life. Direct hernia is more common in elderly people.
2. Sex: Common in males.
3. Symptoms:
 - History of swellings in the groin – *inguinal swelling* which is gradually increasing in size. The swelling becomes more prominent on standing or straining and gets smaller or disappears on lying or on manipulation, unless it is complicated.
 - The swelling may expand and extend down to the scrotum – *inguinoscrotal swelling*.
 - Local discomfort or pain which gets worsened on strenuous effort. The pain may be aching sensation in the groin due to the stretching of the deep inguinal ring by the protruding viscus.
 - The pain may be dragging indicating omentocele. As the omentum is attached to stomach and supplied by T12 nerve, the pain may be referred to the umbilicus.
 - The swelling may be asymptomatic – the hernia may be accidentally discovered during a medical checkup.
 - Sudden severe pain in the hernia with vomiting and irreducibility indicates obstruction of the hernia.
 - History of chronic strain e.g., cough, constipation and difficulty in passing urine should be enquired as these may be the cause of hernia formation.
4. Shape: A bubonocoele or direct hernia is globular in shape. A funicular or vaginal hernia is usually pyriform in shape.
5. *Expansile impulse on coughing* – visible and palpable: present.
6. Non-tender if uncomplicated.
7. Consistency – depends on the contents of the sac: soft, elastic and fluctuant, resonant on percussion – if it contains gut.
Firm and doughy, non-fluctuant and dull on percussion – if it contains omentum.
8. **Getting above the swelling in presence of inguinoscrotal swelling.**
 - To differentiate a scrotal swelling from inguinoscrotal swelling.
 - With the patient standing, normally the spermatic cord is palpated between the thumb and the fingers at the root of the scrotum.
 - (i) In case of scrotal swelling e.g., hydrocele, the spermatic cord can also be felt easily, above the swelling – *so, getting above the swelling is possible.*
 - (ii) In inguinoscrotal swelling, funicular and complete indirect hernia, the spermatic cord cannot be felt nakedly, because it is covered anterolaterally by the sac, *so getting above the swelling is not possible.* On complete reduction of swelling, one can feel the cord at the root of the scrotum.
9. **Relation of the sac with pubic tubercle:** To differentiate inguinal from femoral hernia.
 - Inguinal hernia – the swelling is situated above and medial to the pubic tubercle.
 - Femoral hernia – the swelling is situated below and lateral to the pubic tubercle.
10. **Reducibility:** All herniae are reducible unless complicated.
 - On lying down – the direct hernia usually reduces immediately and spontaneously, but indirect hernia usually requires manipulation.
 - A direct hernia reduces directly backwards.
 - An indirect hernia reduces in an upward, backward and lateral direction.
 - During reduction confirm the contents of the sac – if there is gurgling sound, it is an enterocele; if the sac feels doughy, it is omentocele.

Examination

The patient should be examined first in the standing position and then in lying down position (supine), if necessary.

1. (a) *Inguinal swelling* – indirect hernia: bubonocoele or direct hernia (Fig. 10.4A).
- (b) *Inguinoscrotal swelling* – indirect hernia: funicular or vaginal hernia (Fig. 10.4B&C).

- In *enterocele*, during reduction the first part of the content is difficult to reduce but the latter part is easy to reduce.
 - In *omentocoele*, during reduction the first part is easy to reduce while the latter part is difficult to reduce.
 - After reduction:
 - Note the relation of the sac with the pubic tubercle.
 - *Note the reappearance of the swelling on straining or coughing:*
 - In indirect hernia, the swelling reappears gradually and steadily in medial and downward directions.
 - In direct hernia, the swelling reappears immediately straightforward exactly where it was observed before.
9. *Deep ring occlusion test:*
- Identification of deep inguinal ring – *it lies 1 cm above the femoral pulse felt below the inguinal ligament.*
 - After the reduction of hernia, the deep ring is occluded by the pressure of the thumb and the patient is asked to cough:
 - Indirect hernia – the swelling does not reappear, so the test is positive.
 - Direct hernia – the swelling reappears immediately, so the test is negative.
10. *Invagination test:* After the hernia is reduced, the tip of the index finger can be invaginated into the neck of the scrotum and then pushed inside the inguinal canal through the superficial inguinal ring – the patient is then asked to cough –
- Indirect hernia – a thrust will be felt at the tip of the finger.
 - Direct hernia – a thrust will be felt at the pulp of the finger.

Invagination test helps to diagnose an early case of incomplete hernia. It also helps to differentiate bubonocoele and direct hernia from femoral hernia.

Invagination test is not a must. When necessary, it should be performed very gently *not to cause any pain to the patient*. This test cannot be done in female as the labial skin is thick and not lax.

- Examine the opposite side also to exclude hernia. Direct hernia is often bilateral.
- Examine the testis, epididymis & spermatic cord.
- Examine the tone of abdominal muscles by head or leg rising test. Also look for Malgaigne bulgings. Valsalva manoeuvre may be used to check the tone of the abdominal muscles.
- Note for any evidence of chronic straining like chronic bronchitis, enlarged prostate (per rectal examination), stricture urethra (palpation of bulbar urethra for thickening).

Differential diagnosis

Depends on whether the swelling is incomplete or complete.

- When the swelling is incomplete, i.e., *inguinal or groin swelling*.
- When the swelling is complete i.e., *inguino-scrotal swelling*.

Differential diagnosis of groin swellings

- Femoral hernia
- Enlarged inguinal lymph nodes
- Saphena varix
- Femoral aneurysm
- Lipoma of the cord
- Encysted hydrocele of the cord
- Undescended or ectopic testis
- Psoas abscess
- Malgaigne's bulge.

Femoral hernia

The globular swelling is below the inguinal ligament and below and lateral to the pubic tubercle. Also the swelling is just medial to the femoral vessels. In inguinal hernia the swelling is above the inguinal ligament and above and medial to the pubic tubercle.

In femoral hernia the inguinal canal remains empty when invagination test through the superficial ring is performed.

Enlarged inguinal lymph nodes

Enlarged lymph nodes are found below the inguinal ligament, usually multiple with irregular consistency. Cough impulse is absent. When tender and inflamed look for a septic focus on his leg, his lower abdomen, or his buttock. If suspected look for evidence of tuberculosis elsewhere.

Saphena varix

The swelling is below the inguinal ligament and associated with varicosities of the long saphenous vein. The swelling is soft and easily compressible (unless it is thrombosed), which fills up again when the pressure is released. On coughing the swelling imparts a fluid thrill. The swelling disappears on elevation of the limb with the patient lying down. On auscultation a venous hum may be heard.

Femoral aneurysm

Swelling is below the inguinal ligament and on palpation there is expansile impulse corresponding with the radial pulse. On auscultation a bruit may be heard.

Encysted hydrocele of the cord

A smooth elongated, tense cystic swelling. Not reducible. Cough impulse absent. On traction of the testis the swelling comes down and becomes fixed. Transillumination is positive.

Lipoma of the cord

Features are the same as those of the hydrocele of the cord but transillumination is negative.

Undescended or ectopic testis

Ipsilateral scrotum is empty. Cough impulse is absent unless there is a coexistent hernia. Characteristic testicular sensation is present on pressure over the swelling.

Psoas abscess

A fluctuant swelling below the inguinal ligament. Cross fluctuation positive when the swelling is associated with iliopsoas abscess. The swelling may be partially reducible but there is no true expansile impulse on coughing. There will be evidence of tuberculosis of the spine or hip on clinical and/or radiological examination.

Malgaigne's bulge

Malgaigne described a bulge in the inguinal region which occurs in old people with poor physique but it has no cough impulse. Bulge is usually bilateral. It may be the precursor of a direct inguinal hernia.

Differential diagnosis of inguinoscrotal swelling

1. Congenital hydrocele
2. Infantile hydrocele
3. Encysted hydrocele of the cord

4. Hydrocele of the canal of Nuck (in female)
5. Lipoma of the cord
6. Lymph varix or lymphangiectasis of the cord.
7. Funiculitis.
8. Varicocele

Infantile hydrocele

The swelling appears as inguinoscrotal swelling. The swelling is fluctuant, also cross fluctuation positive (*bilocular hydrocele*), translucent and irreducible. Cough impulse is absent.

Varicocele

Presence of characteristic feel like 'Bag of worms'. On coughing – it imparts a fluid thrill. On lying down when the scrotum is elevated the swelling reduces gradually and disappears. On standing the swelling reappears – fills up from the bottom of the scrotum, whereas the hernia descends from above and the descent can be prevented on deep ring occlusion test.

Funiculitis

A thickened oedematous, tender spermatic cord can be felt with no cough impulse.

Congenital hydrocele

Occurs commonly in children. The swelling is fluctuant and translucent. It reduces very slowly.

Encysted hydrocele of the cord

Already discussed.

Treatment of inguinal hernia

- Conservative
- Operative

Conservative***A. No treatment***

In a patient with severe medical comorbidities with a short life expectancy.

Old patients with chronic bronchitis should not be considered unsuitable for operation because they are in danger of getting herniae strangulated. With modern anaesthesia surgery can be undertaken safely in all ages. If necessary the operation can be performed under local anaesthesia.

B. Truss treatment

A truss never cures a hernia. It is an appliance which is designed to prevent descent of the contents into the sac.

Differences between indirect and direct inguinal herniae

Points	Indirect	Direct
• Age	At any age but common in infants and young adults.	Usually in the elderly persons.
• Site	Usually unilateral (30% bilateral).	Usually bilateral.
• Extent	May be incomplete or complete.	Usually incomplete.
• Entrance	Through the deep inguinal ring.	Through the floor of the Hesselbach's triangle.
• Exit	May be through the superficial inguinal ring. Can descend into the scrotum.	Rarely through the superficial inguinal ring. Does not descend into the scrotum.
• Relation with the inferior epigastric vessels	Inferior epigastric vessels lie close to the medial site of the neck of the sac.	Inferior epigastric vessels lie lateral to the neck of the sac.
• Relation with the spermatic cord	The sac lies within the coverings of the spermatic cord. The sac is anterior to the cord.	The sac lies outside the coverings of the spermatic cord. The sac is either behind, below or above the cord.
• Shape of the swelling	Usually pyriform shape, with the narrow end above and the broad base below.	Usually globular with the broad base above.
• Appearance of the swelling	Slowly appears on straining or coughing.	Easily pops out on standing, straining or coughing.
• Reducibility	Gradually reduces, may need manipulation.	Usually reduces immediately and spontaneously on lying down.
• Direction of reduction	Reduces upwards, then laterally and backwards.	Reduces upwards and backwards.
• Deep ring occlusion test after reduction	Positive, i.e., the swelling does not appear when the patient coughs.	Negative i.e., the swelling reappears.
• Release of occlusion followed by coughing	The swelling appears first in the middle of inguinal region and then flows medially and ultimately turns down to the neck of the scrotum.	The swelling reappears straight forward exactly where it was before.
• Obstruction/strangulation	Common due to narrow neck of the sac.	Rare due to wide neck of the sac.

Differences between enterocele and omentocoele

Points	Enterocele (presence of intestine in the sac)	Omentocoele (presence of omentum in the sac)
Inspection	Visible peristalsis is usually seen.	Peristalsis is never seen.
Palpation	Consistence is soft, elastic and fluctuant.	Consistence is firm, doughy and non-fluctuant.
Percussion	Usually resonant on percussion.	Dull on percussion.
Reducibility	Reduction is easy. The first part is difficult to reduce than the last part which slips in easily. Gurgling sounds can be heard during reduction.	Reduction is usually difficult. The first part reduces easily but the last part resents to be reduced. No gurgling sound is heard.
Auscultation	One may hear peristaltic sound.	No peristaltic sound can be heard.

Mode of action

- The truss acts by pressing the anterior wall against the posterior wall of the inguinal canal. It also compresses the deep inguinal ring.
- By repeated friction it causes adhesion of the

sac with the wall of the canal. This might cause difficulty during operation when necessary.

Indication

- In infants: A truss often helps in spontaneous cure. But when the hernia is associated with undescended

testis a truss should never be used, rather early operation is indicated.

- In old patients: When surgery is contraindicated because of associated general disease, e.g., severe cardio-respiratory disorder.
- Those who refuse treatment: Truss may be advised on request only, but they must be informed about the problem of truss.

Contraindications

- Irreducible hernia.
- Patients with source of chronic strain, e.g., chronic constipation, prostatism, chronic bronchitis; patient doing strenuous job.
- Patients with poor intelligence and perseverance as the wearing of the truss needs careful attention to its positioning and to cleanliness in the area.
- Hernia with huge hydrocele.
- Hernia associated with undescended testis.

Problems of truss

- Improper use can lead to obstruction or strangulation of the hernia. This occurs frequently to patient with poor intelligence or with a persistent source of chronic strain.
- Improper cleanliness causes unhealthy skin which may result in poor wound healing when operation is necessary.
- Prolonged use causes attenuation of the musculature which interferes with subsequent repair and this might be a factor for failure of operation.
- Adhesion due to repeated frictions can cause difficulty during operation when necessary.

For these disadvantages operation should be insisted upon whenever possible.

Types of truss

'Rat-tailed' or 'Adder-headed' truss. The truss must fit snugly over the inguinal canal.

Method of use

- The truss should be applied in lying down position after reduction of hernia completely.
- It should be used constantly except when the patient is in bed. It should be worn again before getting up from the bed.
- The skin covered by the pad and the perineum should be kept clean by daily toilet.

C. Taxis

It implies vigorous manipulation in an attempt to reduce an acute obstructed hernia of short duration only but without any feature of intestinal obstruction or strangulation.

Procedure

- Should be done by an experienced surgeon.
- Should be done after admitting the patient in the hospital.
- Patient should lie down supine on a warm bed with the foot end of the bed raised by 9 inches block.
- Immediate IV/IM injection of pethidine is given to allay the pain and stress.
- Wait for 20-30 minutes. Then IV Calmpose and IV Baralgan/Buscopan is given.
- Once the patient is well sedated, manipulation is tried to reduce the hernia.
- Patient should be observed for 24 to 48 hours, and examined repeatedly to exclude recurrence of hernia, and/or development of intestinal obstruction or strangulation.
- During this observation, patient is allowed plain water and electrolytes orally only. If necessary IV elementation and NPM are advised.
- Once the hernia remains in reduced position, plan for an elective operation.

Dangers of taxis

Rarely attempted now-a-days because of dangers:

- (i) *Reduction-en-masse* – the sac together with its contents gets pushed back into the abdomen, without actually the contents being pushed out of the sac. As a result, constriction around the neck of the sac persists leading to obstruction and strangulation.
- (ii) Reduction of contents into a loculus of the sac, so persistence of obstruction.
- (iii) Contusion or rupture of the intestine contained within the sac.
- (iv) Extraperitoneal reduction – the sac ruptures leading to reduction of contents extraperitoneally.

Operative

Operation is the treatment of choice and it should be advised whenever possible.

- In indirect hernia, the elective operation should be performed as early as possible because chances of obstruction or strangulation are more common with it due to its narrow neck.
- Elective repair under controlled circumstances is far safer with low morbidity than to wait and then operate urgently in the face of the potential serious complications of bowel obstruction or strangulation.
- In direct hernia, as the chances of obstruction or strangulation are much less common due to its wide neck, one can delay the trial of operation.
- Elderly men with asymptomatic or minimal symptoms of inguinal hernia may also undergo a period of observation.
- In an uncomplicated hernia, the surgeon should treat first any chronic source of strain from constipation, prostatism, stricture urethra or chronic cough before an operation. In presence of any chronic strain the repair of hernia very often fails and results in recurrence. Correct chronic cough with stoppage of smoking, postural drainage, chest physiotherapy and antibiotics as necessary. Correct chronic constipation, prostatism or structure urethra first.
- Operation can be performed under local, regional, epidural anaesthesia or general anaesthesia.
- *Infant of only few days old: Wait until the baby is 6 months old.* Obstructions unlikely in infancy because the neck of the sac is fairly wide and the inguinal canal is straight and short.

Operative procedures

Basically three types of operations are available:

1. Herniotomy alone
2. Herniotomy + Herniorrhaphy
3. Herniotomy + Hernioplasty.

Suitability of any one of these methods depends on the age of the patient, type of hernia and the conditions of the musculature of the inguinal canal.

Herniotomy

Indications

1. In infants and children.
2. In adolescents and young adults with good musculature.

3. Prior to herniorrhaphy or hernioplasty in other ages of patients.

It consists of excision of the sac only. The sac is isolated, opened and the contents are reduced. The neck of the sac is twisted and is transfixed and ligated high up near the deep ring followed by excision of the remaining distal portion of the sac.

However, it is to be noted that herniotomy is not performed in direct inguinal hernia; instead the sac is inverted here. In case of a large sac, after the inversion of the sac, the fascia transversalis may be plicated or imbricated to keep the sac reduced.

In infants and children, it is not necessary to open the inguinal canal, as the internal and external rings are superimposed.

Herniorrhaphy

It consists of

1. Herniotomy +
2. *Reconstruction of the posterior wall of inguinal canal* by approximating the conjoined muscle and tendon to the recurved edge of the inguinal ligament with non-absorbable suture such as silk, nylon or prolene. This is known as *Bassini's repair*. Silk is better avoided; if it becomes infected, it will lead to sinuses.

The aim of Bassini's repair is to strengthen the posterior wall of the inguinal canal by pulling down the conjoined muscle and tendon behind the spermatic cord. Interrupted or continuous sutures are applied without tension as this will lead to cutting of the muscles and/or ligaments from the sutures.

Many modifications added to the Bassini's repair have been advocated to strengthen the repair further:

- (a) To avoid tension, one may add *Tanner's slide operation*: Here a curved release incision is made on the anterior rectus sheath above the conjoined tendon so that the lower lateral leaf of the sheath at once retracts down and makes the conjoined tendon loose.
- (b) *Lytle's repair in indirect hernia*: Tightening of the stretched internal inguinal ring on its medial side if it is too wide. The ring should not be tightened too much. The tip of the haemostat should slide easily.

- (c) Lateralization of the ligated neck of the sac after excision.
- (d) Plication of fascia transversalis: in direct hernia.
- (e) *McVay's repair* involves suturing of the conjoined tendon to the Cooper's ligament medial to the femoral vein and to the ilio-pubic tract, above and lateral to the femoral vein. Because the femoral ring is also closed during this procedure this is a good method of repair when an associated femoral hernia is noted during repair of an inguinal hernia. However, the chances of femoral vein injury are common.
- (f) *Shouldice repair*: It is a modification of Bassini's repair developed at the Shouldice clinic in Toronto. It is basically a multilayered (Four layers) Bassini's repair.
 - First two layers involve double breasting of the fascia transversalis.
 - 3rd layer involves suturing of the conjoined tendon to the inguinal ligament.
 - 4th layer involves suturing the anterior rectus sheath and the conjoined tendon to the inner surface of lower leaf of external oblique muscle.
- (g) *Halstead modifications*: Here the spermatic cord is exteriorised by suturing both the flaps of external oblique aponeurosis behind it, thus the cord remains in subcutaneous plane.
- (h) *Willy-Andrew's modifications*: Here the upper flap of external oblique aponeurosis is brought down behind the cord and is sutured to the inguinal ligament and the lower flap is brought over the cord and sutured to the upper flap, so that the cord remains sandwiched between the two flaps of external oblique.

Indication: In middle aged or elderly patient with weak abdominal musculature.
- (i) *Kuntz operation*: In elderly patient with large scrotal hernia and weak abdominal musculature Kuntz operation is advocated. Here orchidectomy along with excision of spermatic cord distal to the deep ring is carried out followed by obliteration of the inguinal canal. This will guard against recurrence. So, in old men permission for orchidectomy should be taken in writing before operation.
- (j) In women, it is advisable to remove the round ligament.

Hernioplasty

Indications

- (a) In elderly patients with grossly weak abdominal musculatures.
- (b) Recurrent hernia.

It consists of

1. Herniotomy +
2. *Reinforcement of the weak posterior wall of the inguinal canal by filling the defect of the posterior wall with some material* – autogenous or heterogenous.

Uses of autogenous materials (i.e., patient's own tissues) are obsolete now.

Heterogenous materials such as prolene, nylon or stainless steel wire, or a prolene or Marlex mesh may be used.

(a) *Darning*: After herniotomy (in indirect hernia) or inversion of the sac (in direct hernia) gap between the conjoined tendon and the inguinal ligament is covered with the prolene suture without tension in criss-cross manner to strengthen the posterior wall. The procedure is called darning. The fibroblasts and capillaries grow in between the interstices of the darn, thus converting it into a thick fibrous sheath.

(b) *Lichtenstein tension-free mesh repair*: The posterior wall of the inguinal canal is covered by a prolene mesh (size 8 cm × 6 cm), in a tension-free manner without closing the gap by direct suturing. The mesh is sutured to the conjoined tendon with transversalis fascia, pubic tubercle, lacunar ligament and inguinal ligament deep to the spermatic cord.

Operation for direct inguinal hernia

Herniotomy is not performed; instead the sac is inverted after reduction of the contents followed by plication of the fascia transversalis. This is then followed by some form of repair of the posterior wall as mentioned in herniorrhaphy or hernioplasty.

Laparoscopic repair of inguinal hernia

Inguinal hernia may be repaired laparoscopically by pre-peritoneal placement of mesh.

Two methods are in wide use:

1. *Totally extra-peritoneal approach* (TEP)
2. *Trans-abdominal preperitoneal approach* (TAPP)

These are based on Stoppa's method of reconstruction of the weakened anterior abdominal wall by preperitoneal placement of the mesh between the posterior surface of the anterior abdominal wall and the peritoneum. The mesh may or may not require any fixation by anchoring sutures or tacker. In absence of fixation, the mesh is kept in place by Pascal's principle of hydrostatic pressure: the intraabdominal pressure acting via the anterior peritoneal layer keeps the mesh steadily against the abdominal wall.

Indications

1. Both indirect and direct inguinal herniae
2. Bilateral inguinal herniae
3. Recurrent hernia.

Advantages

1. Laparoscopic mesh repair also takes consideration of coverage of femoral ring and obturator ring in addition to direct and indirect inguinal rings.
2. Patient can be discharged within 48 hours.
3. Early return to work.

Complications of hernia operation

Complications during the operation:

1. Injury to the external iliac or femoral vessels.
2. Injury to the vas deferens especially in children.
3. Injury to the urinary bladder and colon especially with sliding hernia.
4. Injury to the inferior epigastric vessels.
5. Injury to the contents of the sac.
6. Injury to the testicular artery.

Early post-operative complications:

1. Retention of urine.
2. Haematoma of the cord and scrotum.
3. Wound infection.

Late post-operative complications:

1. Recurrence.
2. Sinuses.
3. Neuralgic pain due to involvement of ilio-inguinal nerve in the suture – causes hyperaesthesia over the medial side of inguinal canal.
4. Painful scar.
5. Atrophy of the testis due to injury to testicular

artery or due to compression of the spermatic cord following narrowing of the internal ring too tight.

6. Osteitis pubis.
7. Mesh extrusion with or without foreign body reaction.
8. Epidermoid cysts – when the skin flap is used for plastic repair.

Causes of recurrence of hernia

The recurrence rate is between 3 and 7 percent. Most of the recurrences occur by the end of first year.

A recurrent hernia may be indirect or direct. The following factors are responsible for the recurrence:

1. Weak abdominal muscles.
2. Persistence of chronic strain leading to chronic raised intra-abdominal pressure, e.g., constipation, prostatism, chronic bronchitis.
3. Presence of both indirect and direct hernial sacs, i.e., saddle hernia but failure to recognise either of them during operation.
4. Failure to dissect the sac right up to the neck and inadequate removal of the sac.
5. Inadequate repair of the posterior wall of the inguinal canal – either too tight or too loose a repair.
6. Inadequate haemostasis.
7. Wound infection.
8. Too early resumption of strenuous labour.

Management of recurrent hernia:

1. Exclude or correct any persistent source of chronic strain, e.g., chronic cough, constipation, straining at micturition. Ultrasound of KUB is useful to exclude enlarged prostate or bladder neck obstruction.
2. Following operations may be helpful.
 - (a) Lichtenstein tension-free mesh repair is preferable.
 - (b) In an elderly patient, Kuntz operation may be added to Lichtenstein repair.
 - (c) Shouldice repair.
 - (d) Stoppa's preperitoneal mesh placement repair (hernioplasty).
 - (e) Laparoscopic repair with pre-peritoneal mesh replacement.

RARE VARIETIES OF INGUINAL HERNIA

Dual Hernia

- Also known as *saddle bag* or *pantaloon hernia*.
- It has two sacs – one direct and another indirect sac lying on either side of the inferior epigastric vessels; both are connected by an isthmus which lies behind the inferior epigastric vessels.

Significance:

1. Deep ring occlusion test may be confusing as it only controls the indirect sac.
2. *One of the causes of recurrence* if one sac, particularly the indirect sac, is missed during operation. *So, during direct hernia repair one should explore the internal ring and the coverings of the spermatic cord to exclude indirect hernia.*

Sliding Hernia

- Also known as *Hernia-en-glissade*.
- It is a type of hernia in which a portion of extra-peritoneal bowel e.g., caecum, colon or urinary bladder, may slide down into the inguinal canal pulling a peritoneal sac with it. Thus the posterior wall of the sac is formed by both the peritoneum of the sac and by the wall of a viscus which lies behind the peritoneum.
- The sac may contain other loops of bowel or omentum. But if the caecum and appendix are the contents of the hernial sac, it is not a sliding hernia.

Clinical features:

1. Most commonly seen in elderly men.
2. Particularly occurs in long-standing cases.
3. Left-sided hernia is more common than right-sided.
4. This hernia is suspected when a large globular hernia descends down into the scrotum.
5. Complete reduction is not possible.
6. After reduction, sliding hernia reappears very slowly.

Significance:

1. This hernia can easily be strangulated. There may be strangulation of small bowel inside the sac and/or strangulation of large bowel forming the wall of the sac.

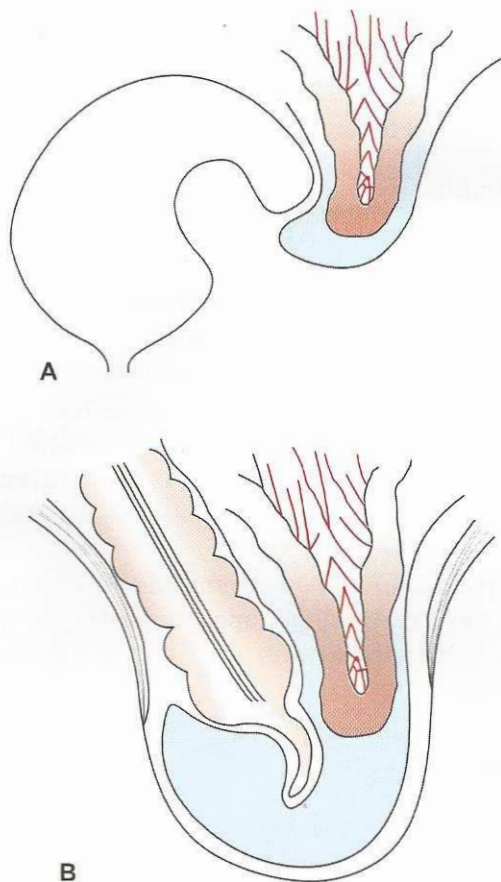


Fig. 10.7. Sliding hernia. A. Posterior wall of the hernial sac is formed by the part of the urinary bladder wall which lies behind the peritoneum of the sac. B. Posterior wall of the hernial sac is formed by the part of the caecal wall which lies behind the peritoneum of the sac.

2. The real danger associated with a sliding hernia is the failure to recognise the visceral component of the hernia sac before injury to the bowel or bladder during operation.

Treatment:

1. Truss treatment is absolutely contraindicated.
2. The hernial sac should be dissected carefully to avoid injury to colon or bladder.
3. Once the hernial sac is opened and the contents are reduced back in the abdomen, the sac should not be twisted to avoid inclusion of colon wall or bladder wall. Instead the distal sac is removed and the proximal part of the sac is closed with a purse-string suture. The hernia repair is then completed. A mesh repair is preferable.

4. In elderly patient, orchidectomy is advised to furnish complete closure of the inguinal canal and thus to avoid recurrence. Hence, prior consent for orchidectomy should be taken.

Interstitial Hernia

- In this hernia, the hernial sac instead of being present in the inguinal canal, may lie in the musculofascial layers of the abdominal wall.
- The hernia is usually incomplete.
- Following varieties may be encountered:
 - (i) *Preperitoneal*—the hernia sac lies between the peritoneum and the fascia transversalis.
 - (ii) *Intraparietal*—the hernia sac lies between the internal oblique and the external oblique aponeurosis.
 - (iii) *Extraparietal*—the hernia sac lies outside the external oblique aponeurosis in the subcutaneous fat.
- *Hernia in association with undescended testis is commonly one of interstitial variety.*

Ogilvie Hernia

- This is a rare variety of direct inguinal hernia.
- Here the hernia sac *protrudes through an oval defect in the conjoined tendon.*
- So it is also known as funicular direct inguinal hernia (cf. funicular indirect inguinal hernia, prevesical hernia).
- Chances of obstruction are common than usual direct hernia.

Prevesical Hernia

- It is also called funicular direct hernia.
- Rare variety. Occurs principally in elderly men.
- The hernia sac containing a portion of bladder wall and prevesical fat protrudes *through a small oval defect in the conjoined tendon* just above the pubic tubercle.
- A history of swelling in the groin becoming less prominent or disappear after micturition is present.
- Operation should always be advised except in patients with significant medical comorbidities.
- Chances of obstruction are more common than usual direct hernia.
- Chances of bladder injury are frequent during operation if not careful.

Richter's Hernia

- In this hernia, only a portion of the circumference of bowel wall becomes protruded in the hernia sac.
- *This condition often complicates a femoral hernia and rarely an obturator hernia.*
- Chances of strangulation without complete obstruction of bowel lumen is common.
- But the diagnosis is frequently delayed, because the patient may present with gastroenteritis i.e. diarrhoea with or without vomiting. The intestinal colic may be present but the bowels open normally.
- Intestinal obstruction may develop late when half of the circumference of the bowel is involved.
- *Often, diagnosis is established after exploratory laparotomy in a case of intestinal obstruction.*

Littre's Hernia

- In this condition Meckel's diverticulum is the content of the sac.

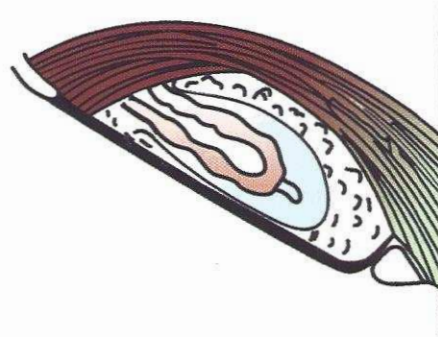


Fig. 10.8. Littre's hernia.

Maydl's Hernia

- Also known as Hernia-en-W.
- In this condition, two adjacent loops of bowel remain in the sac, the connecting portion remains inside the abdomen. The bowel loops in the hernial sac looks like W.
- This connecting portion of W loop is more vulnerable to become strangulated early in an obstructed hernia. Thus the strangulated piece of the gut remains inside the abdomen (Fig. 10.9).

Clinical features:

1. The patient presents with obstructed hernia with intestinal obstruction.

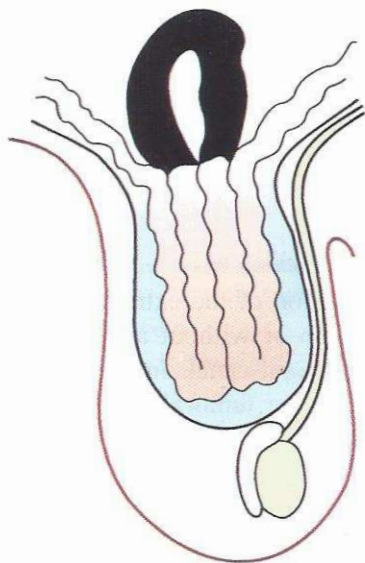


Fig. 10.9. Maydl's hernia.

2. On examination, there are no signs of gangrene over the scrotum and groin, but tenderness and rebound tenderness will be elicited over the lower abdomen above the inguinal ligament.

On exploration: After examining the bowels inside the sac, the intra-abdominal segment of bowel is to be examined for gangrene and the gangrenous part should be excised.

CONGENITAL INGUINAL HERNIA

- Develops due to failure of obliteration of processus vaginalis.
- *Always indirect type. Usually appears as an inguinoscrotal swelling since birth.* It gradually increases in size.
- The swelling appears on straining or crying, on crawling, sitting and standing.
- The swelling disappears when the child lies down.
- The swelling may transilluminate (cf. congenital hydrocele).
- Reducible with or without gurgling sound.
- The swelling will reappear, if the child is induced to cry.
- In newborn infant, the inguinal canal is not developed and the deep and superficial inguinal rings lie close or superimposed to each other.

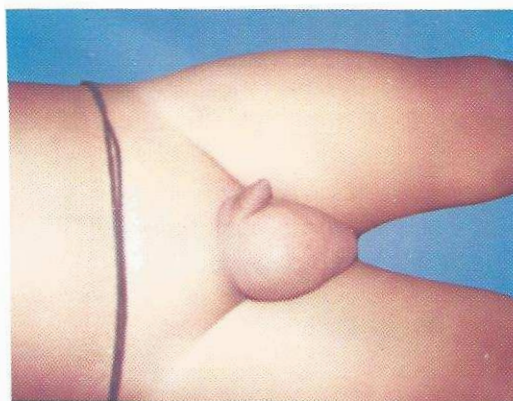


Fig. 10.10A. Congenital right inguinal hernia.



Fig. 10.10B. Congenital right inguinal hernia – presented with intestinal obstruction. Note the dilated small bowel loops right upper abdomen – step ladder pattern.

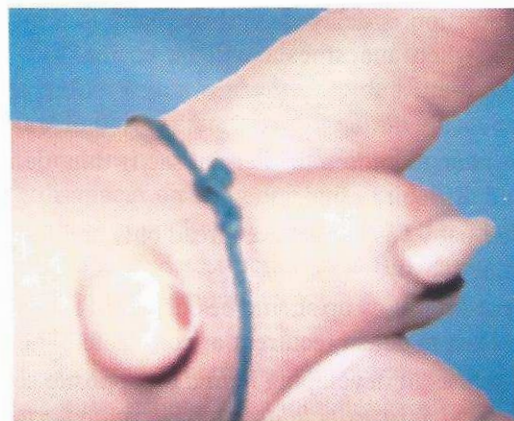


Fig. 10.11. Congenital umbilical hernia with left inguinoscrotal hernia.

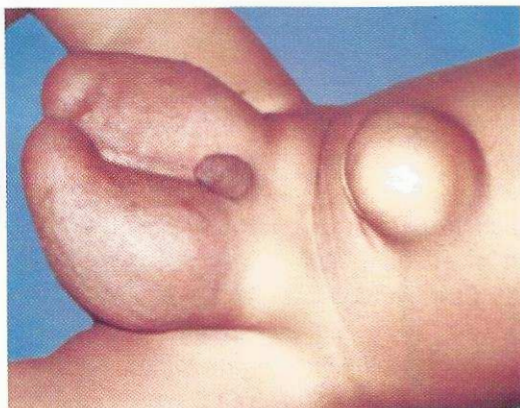


Fig. 10.12. Congenital umbilical hernia and bilateral inguinal herniae. Both are reducible.

- By the age of 2 years, as the deep ring moves laterally, the inguinal canal is formed with wide separation of the two rings.
- *Early operation is advocated in infants and children* because of high chance of obstruction though strangulation is rare.
- *Only simple herniotomy is required.* No herniorrhaphy is attempted. Because the inguinal canal is short and straight and the external ring lies directly over the internal ring, opening of inguinal canal is not required.

Problems of inguinal hernia in children:

- It is very difficult to distinguish congenital hernia from a congenital hydrocele because they both transmit light of a torch. The distinction is not critical because most hydroceles require operation.
- Operation is delayed until the baby is 6 months old when anaesthesia will be easier. In a rare fortunate baby, his hernia may resolve spontaneously during this 6 months period of waiting.
- In young children the hernial sac is thin, delicate and difficult to find.
- The baby vas is thin like thread only and liable to injury easily.
- Post-op follow up should be continued for 5 years at least to detect recurrent or contralateral hernia.
- Chances of developing hernia on opposite side—*Around 5-10% of children may develop hernia on contralateral side during follow up.*

- Routine contralateral exploration which was advocated earlier is not advised now-a-days.
- Laparoscopic suturing of the internal ring is a less traumatic operation. Moreover, *both the internal rings are inspected from inside at the same time and tackled accordingly at the same sitting.*

FEMORAL HERNIA

It is the protrusion of the extraperitoneal fat and peritoneum with or without abdominal contents through the femoral canal. In this hernia the sac passes through the femoral ring and enters the femoral canal.

Surgical anatomy

Femoral sheath: This is a funnel shaped fascial prolongation around the femoral vessels below the medial half of the inguinal ligament. The anterior wall of the sheath is formed by the fascia transversalis and the posterior wall by the fascia iliaca. The sheath has got three compartments separated by fibrous septae:

1. Lateral compartment — contains femoral artery and femoral branch of genitofemoral nerve.
2. Intermediate compartment — contains femoral vein.
3. Medial compartment — femoral canal.

Femoral canal: It is the innermost compartment of the femoral sheath. The canal is about 2 cm long and is conical in shape with the base directed upwards and the apex directed downwards. At the base lies

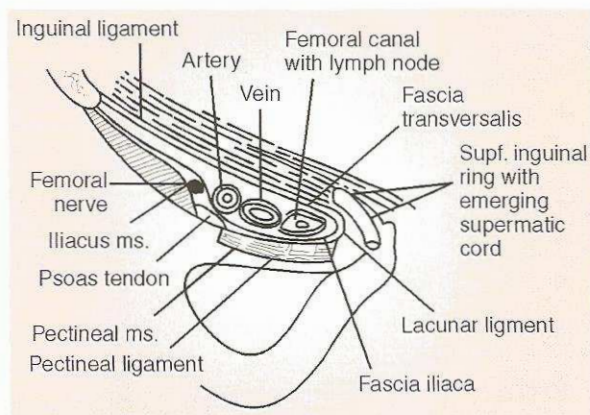


Fig. 10.13. Femoral canal – its boundaries and relations.

the femoral ring which is the mouth of the canal. The canal extends from the femoral ring above up to the saphenous ring below.

Boundaries of the femoral ring

- *Anteriorly*: Inguinal ligament.
- *Posteriorly*: Pectineal line of the horizontal ramus of the pubic and the pectineal ligament.
- *Medially*: Crescentic edge of the lacunar ligament.
- *Laterally*: Fibrous septum separating the canal from the femoral vein.

All these boundaries are rigid except the fibrous septum laterally.

The ring is closed above by the septum crurale — a condensed extra-peritoneal tissue pierced by the lymphatic vessels.

In 20% cases the obturator artery is replaced by the pubic branch of the inferior epigastric artery which, when present, courses along the crescentic margin of the lacunar ligament. *This abnormal artery is vulnerable* — if the lacunar ligament needs division for release of the constricted ring for reduction of the femoral hernia.

Contents of the femoral canal

1. Fibro-fatty tissue.
2. Lymph nodes and lymphatics. Lymph node situated at the ring is known as Cloquet's node.

Surgical pathology

1. Majority of femoral herniae are acquired in nature.
2. Femoral herniae are *more common in females*. *More common in the middle aged and the elderly females* due to wide pelvis and repeated pregnancy. Repeated pregnancy causes increased abdominal pressure which is probably an initiating factor. However, inguinal hernia is more common than femoral hernia in females. Femoral hernia is almost unknown in infants.
3. *Right-sided hernia is more common* than the left-sided one. The hernia may be bilateral.
4. *Course of the hernia*: The hernial sac enters through the femoral ring and descends vertically down along the femoral canal up to the lower border of fossa ovalis where it lies in close relation to the long saphenous vein. Due to

attachment of the superficial fascia of the abdomen (fascia of Scarpa) to the fascia lata of the thigh at the lower border of the fossa ovalis, the sac cannot pass down in the thigh. So the sac courses forward, and when enlarging it courses upward over the inguinal ligament and external oblique aponeurosis and *thus the distal part of the sac overlies the inguinal ligament and occupies the inguinal region*. The shape of the sac then becomes retort-shaped.

Contents of the sac

May contain:

1. A part of the omentum.
2. A loop of small bowel.
3. A part of the circumference of small bowel (Richter's Hernia).

Coverings of the sac

From superficial to deep:

1. Skin.
2. Superficial fascia.
3. Cribriform fascia covering the saphenous opening.
4. Fascia transversalis — representing the anterior femoral sheath.
5. Extraperitoneal fat.
6. Peritoneum — the hernial sac.

Diagnostic criteria

1. *Age*: Middle-aged or elderly persons commonly.
2. *Sex*: More common in females.
3. *Symptoms*:
 - The patient complains of dragging pain in the groin.
 - Usually she or he presents with a small globular swelling in the groin. It becomes more prominent on standing or straining but may disappear on lying down.
 - An obese lady may present with the features of intestinal obstruction or strangulation of an unnoticed femoral hernia.

Examination:

- The femoral hernia should be examined in the similar manner as in inguinal hernia. Ask the patient to stand up, note the exact position of the lump, *try to determine the exact anatomical relations of the lump to the inguinal ligament*

Differences between inguinal and femoral herniae

Points	Inguinal	Femoral
• Age	Any age	Middle-aged or elderly persons commonly
• Sex	More common in males	More common in females
• Side	Unilateral; common on the right side	Unilateral; common on the right side
• Relation with the pubic tubercle	The sac lies above and medial to pubic tubercle	The sac lies below and lateral to pubic tubercle
• Cough impulse	Usually present	Usually absent
• Reducibility	Usually reducible; the hernia reduces in an upward, backward and lateral direction	Often partial; when complete, the hernia reduces directly upward and backwards
• Invagination test after reduction	On coughing by the patient: a thrust will be felt at the tip of the finger	On coughing: no thrust will be felt; the inguinal canal found to be empty

and pubic tubercle and then note if the lump has a cough impulse and whether it is reducible.

4. **Position:** A round or oval swelling below the medial end of the inguinal ligament – at or below the groin crease.
5. **Palpation:** The neck of the lump is situated below and lateral to the pubic tubercle (cf. Inguinal hernia).
6. **Cough impulse:** Both visible and palpable, may be absent in many femoral hernia because the contents are adherent to the peritoneal sac.
7. **Reducibility:** Often partial. When complete it reduces directly upwards and backwards.
8. **Inguinal canal examination, e.g., invagination test – empty.**

Complications

1. Irreducibility
2. Obstruction
3. Strangulation: Femoral hernias have the highest incidence of strangulation (15%) of all hernias.

These are commoner in femoral hernia than in inguinal hernia or any other hernia because the neck of the femoral canal is narrow & has a sharp medial border.

Differential diagnosis

1. Incomplete inguinal hernia or bubonocoele
2. Saphena varix
3. Enlarged lymph node
4. Lipoma
5. Aneurysm of the femoral artery
6. Psoas abscess
7. Rupture of the adductor longus muscle.

Incomplete inguinal hernia

The swelling is in the inguinal region — above the inguinal ligament, *above and medial to the pubic tubercle*. Invagination test through the superficial ring will confirm that the swelling is inside the inguinal canal. The swelling has a cough impulse and is usually reducible.

Enlarged lymph node

An enlarged deep inguinal lymph node may be almost always impossible to distinguish from a femoral hernia. If with antibiotics and rest the swelling does not get smaller it should be explored. If suspicion of strangulated hernia arises — operation should be done immediately without delay.

Lipoma

It is a soft lobulated swelling and mobile. It can never be reducible. Cough impulse absent. If suspicion arises, it should always be explored.

Rupture of the adductor longus muscle

Common in athletes, so history of injury is present. Patient presents with an oval swelling on the medial side of the femoral triangle. Cough impulse absent. Reducibility absent. When the patient is asked to adduct the thigh against resistance — the swelling becomes more prominent and well-defined.

Others

See under D/D of Inguinal hernia (page 170).

Treatment

Always operative.

Conservative treatment has no role in femoral hernia because:

- no truss can be fitted to control the femoral ring;
- risk of obstruction or strangulation is very common in femoral hernia.

Operations

Essentials of operation:

- Herniotomy + closure of the femoral ring either by suturing the inguinal ligament to the pectineal ligament, or the conjoint tendon to the pectineal ligament.

There are three avenues of approach to the femoral hernia:

1. Low or sub-inguinal – Lockwood's operation.
2. Inguinal – Lotheissen's operation.
3. High or supra-inguinal – McEvedy's operation.

Lockwood's operation

Advantage:

A direct approach to the swelling and hence the sac. Speedy and simple method, so suitable for small and uncomplicated femoral hernia.

Disadvantage:

- Difficult to resect a gangrenous bowel so not suitable for the operation of a strangulated hernia.
- Difficult to repair the femoral ring.

Lotheissen's operation

Advantage:

Easy to deal with gangrenous bowel.

Disadvantage:

It causes weakening of the inguinal canal and its posterior wall.

McEvedy's operation

Advantage:

- Easy to deal with gangrenous bowel and to repair the femoral ring.
- It does not damage the inguinal canal.

Disadvantage:

Access to the fundus of the sac is not sufficient, so removal is difficult.

RARE TYPES OF FEMORAL HERNIA

1. Prevascular hernia (Narath hernia)

- In this case, the femoral hernia sac lies behind the femoral vessels.
- Occurs in patient with congenital dislocation of hip

2. Lacunar hernia (Langier's hernia)

In this case, the femoral hernia sac passes through a small defect in the lacunar ligament.

3. Pectineal hernia (Cloquet's hernia)

The femoral hernia sac passes between the pectineal muscle and fascia, behind the femoral vessels.

4. External femoral hernia (Hasselbach's hernia)

The femoral hernial sac lies lateral to the femoral artery.

INCISIONAL HERNIA

Syn.: Postoperative hernia, Ventral hernia

An abdominal incisional hernia is one where the peritoneal sac herniates through an acquired scar in the abdominal wall, usually caused by a previous surgical operation or an accidental trauma. Scar tissue is inelastic and can be stretched easily if subjected to constant strain.

Contents of such hernia are usually bowel and/or omentum.

Pre-disposing factors

1. Some subjects:

- (a) Obese individuals with poor musculature.
- (b) Diabetic patients.
- (c) Malnutrition:
 - Patients with severe anaemia.
 - Patients with hypoproteinaemia.
 - Patients with avitaminosis, e.g., vitamin C deficiency.
 - Patients with cachexia and advanced malignant disease.

- (d) Patients with any chronic source of straining, e.g., chronic cough, enlarged prostate, constipation, or any other factor which leads to increase in intra-abdominal pressure, e.g., ascites.

2. Some operations:

- (a) Operation for peptic perforation or appendicular perforation.
- (b) Operation in case of peritonitis.
- (c) Operation for visceral cancer.
- (d) Pancreatic resection operation.

- (e) Operation in a case of bowel obstruction where decompression of the grossly distended bowel is not properly done.
3. *Some incisions:*
- (a) Kocher's subcostal incision for cholecystectomy causing injury to the ninth and tenth thoracic nerves.
 - (b) McBurney's incision for appendicectomy causing injury to the ilio-inguinal nerve.
 - (c) Battle's para-rectal incision for appendicectomy.
 - (d) Midline infraumbilical incision e.g., during caesarean section.
4. *Some faults during operation:*
- (a) Division of motor nerves supplying the muscles around the wound.
 - (b) Defective closure of the laparotomy wound, e.g., failure of suturing the abdominal wounds in anatomical layers; suturing under tension causes pressure necrosis of intervening tissues.
 - (c) Inadequate haemostasis during operation leading to wound haematoma which becomes more vulnerable if infected.
 - (d) Wound closed with non-absorbable suture materials like nylon, prolene are followed by a lesser incidence of incisional hernia than the wound closed with absorbable suture like catgut.
 - (e) Drainage tubes brought through the main laparotomy wound and left behind for a long time. (Intraperitoneal drains should always be brought out through a separate stab incision.)
5. *Some post-operative complications:*
- (a) Severe post-operative abdominal distensions – causing tension over the suture line.
 - (b) Persistent postoperative cough – causing tension over the suture line.
 - (c) Post-operative peritonitis – because there is a chance of wound infection and abdominal distension.
 - (d) Wound infection.
 - (e) Wound haematoma.
 - (f) Too early removal of sutures.
 - (g) Too early resumption of strenuous labour.

- (h) Steroid therapy in post-operative period.
- (i) Persistence of diabetes, vitamin C deficiency, hypoproteinaemia, etc.
- (j) Failure of proper tissue healing due to abnormal collagen production and maintenance causes weak scar tissue which may lead to late development of incisional hernia after 5 to 10 years of operation.

Pathology

Can be discussed under two headings:

- A. Without infection.
- B. With infection.

A. Without infection

There is a partial and quiet disruption of the deeper layers of a laparotomy wound occurring immediately after operation or during very early post-operative period; but such an event usually passes unnoticed because the skin edges remain apposed with intact sutures.

But during this event sometimes there is oozing of the serosanguineous discharge through the laparotomy wound, when it should be regarded as a *red signal of wound dehiscence or burst abdomen*. This signal demands immediate examination of the laparotomy wound and if necessary resuture of the deeper disrupted layers of the wound.

Following this event there is separation of both sheath and muscles resulting in healing by weak scar tissue.

B. With infection

Wound suppuration – disruptions of the sutures – separations of sheath and muscles – healing by weak scar tissue.

In both cases herniation occurs through this weak scar tissue.

Structural changes

1. Skin over the hernial protuberance is thinned out, stretched and often shows a wide irregular scar. The scar is devoid of subcutaneous fat and is very often adherent to peritoneal sac.
2. The sac is usually big and multiloculated.
3. Contents: The bowel and/or the omentum which are often matted together and may be adherent to the loculated peritoneal sac.

Types of defect

Incisional hernia may be of two types:

1. One type occurs through the midline upper abdominal incision or midline lower abdominal incision where the muscular defect is wide with smooth and regular margins. More frequently this type of hernia takes the form of a diffuse bulge along the whole length of the incision and usually reduces spontaneously as soon as the patient lies down. With this type the risk of strangulation is remote.
2. In the second type, the hernia occurs mainly through the defect in the lateral part of the abdomen where the muscular defect is relatively small and irregular; there may be presence of two or more such defects in the same scar. Risk of strangulation is very common with this type.
3. Size of the defect:
 - Large and wide defect
 - Small defect
 - Multiple defects

Why the midline infraumbilical abdominal incision is frequently followed by an incisional hernia?

Because of the following reasons:

1. The posterior rectus sheath is complete only up to a point midway between umbilicus and symphysis pubis, where it ends in a free border, the linea semicircularis. Below this level the sheath is deficient because all the three aponeurosis pass in front of the rectus muscle. The rectus abdominis muscle here is separated from the peritoneum by the fascia transversalis alone.
2. Below the linea semicircularis the linea alba is thin and narrow and it lies mainly in front of the recti muscles as the posterior sheath is deficient.

So, midline infraumbilical incision results in a weakly supported scar which is likely to be followed by hernia.

Diagnostic criteria

1. The patient usually presents with a bulging in the vicinity of an incisional scar.
2. The bulging may be either localised occurring through a small portion of the scar, mostly in

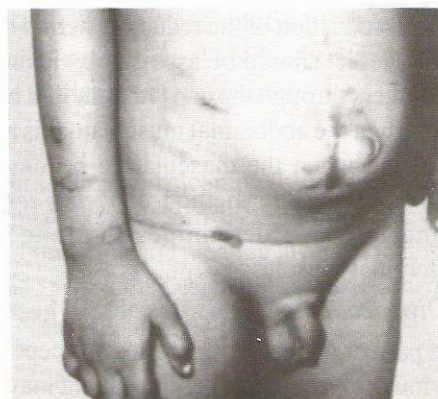


Fig. 10.14. Incisional hernia. Note the swelling with overlying scar. This hernia was reducible. Cough impulse positive.



Fig. 10.15. Large incisional hernia. Note the overlying stretched skin, depigmented irregular scar, ulcers and pigmented areas. This hernia was reducible. Cough impulse positive.

the lower end, or diffuse along the whole length of the scar.

3. The bulge becomes more marked on standing and coughing. Usually it reduces spontaneously as soon as the patient lies down. The reducibility may be partial or complete. Very often the hernia may be completely irreducible.
4. *Expansile impulse on coughing is present.*
5. The skin over the hernia is thin and atrophic and sometimes through this atrophic skin the peristaltic wave of the small intestine may be visible. The overlying skin may be unhealthy, infected or ulcerated.
6. *On palpation*, the edge of the defect can be delineated.

After reduction of the reducible hernia the size of the defect should be assessed by insinuating the fingers through the gap – this should be done both when the abdominal musculature is relaxed fully and when the musculature becomes taut after applying the method of *rising test*. The rising test also determines the tone of the abdominal muscles.

Divarication of the recti is also to be noted.

7. The patient may be symptomless, except for the obvious bulge. Sometimes the patient may suffer from recurrent attacks of subacute intestinal obstruction, or strangulation when the neck of the sac is narrow.
8. History of previous operation, stormy post-operative period, discharge through the wound requiring prolonged dressing etc., is present.

Complications

1. Often patient complains of *discomfort or pain around the scar of the hernia*, mostly after strenuous jobs. The pain is dragging in nature and due to stretching of the gut or omentum adherent to the sac underneath the scar. Often, this pain may incapacitate the patient from his/her regular jobs; so the patient seeks early consultation of the doctor.
2. *Irreducibility*.
3. *Obstruction*: Adhesions of the contents, bowel and omentum, with the sac may lead to angulation and subacute obstruction.
4. *Strangulation*, particularly when the neck of the sac is narrow.

Treatment

- Preventive
- Palliative
- Operative

Preventive

The incidence of incisional hernia can be reduced by eliminating, where possible, the predisposing factors mentioned above.

Post-operatively, convalescence should be gradual and all sorts of strain be avoided. Heavy weight lifting or any strenuous work should be avoided for at least 3 months. The wound must be checked one month

after operation, and any suspected weakness is supported by abdominal corset for a few months.

Palliative

By using abdominal corset fitted with suitable pad, providing that there are no complications within the hernia.

Although trusses are generally not successful, the patient with a large ventral hernia where surgical repair is delayed for medical reasons, or may be difficult, can be successfully managed non-operatively with properly fitted corset and close follow up.

Operative

Indications:

1. Discomfort in the hernia.
2. Hernia with complications such as irreducibility, intermittent attacks of subacute obstruction or complete obstruction.
3. Cosmetic reasons.

Pre-operative measures:

1. Correction of obesity and weight reduction by proper controlled diet.
2. Correction of any other contributory factor as already mentioned (e.g., cough, anaemia, hypoproteinaemia, diabetes, etc.).
3. *Induction of artificial pneumoperitoneum*: In patients with very large ventral hernia containing nearly half the abdominal content, replacement of the contents within the abdominal cavity, is either impossible, or dangerous as it will raise the intra-abdominal pressure greatly (abdominal compartment syndrome) to cause elevation of the diaphragm and thereby respiratory embarrassment, compression of inferior vena cava and venous congestion, paralytic ileus due to mesenteric oedema followed by stasis in the splanchnic bed leading to shock. Any repair under such excessive tension is also likely to give way. To avoid these, it is advisable to prepare these patients by producing artificial pneumoperitoneum at frequent intervals and with increasing pressure to stretch the abdominal wall gradually to increase the abdominal capacity.

Method of repair:

The following methods of operative repair are available, which can be remembered by the mnemonics 'MILK'.

- M** 1. Repair by Mayo's technique (double-breast technique). (see page 192)
- 2. Repair by muscle pedicle flap.
- I** 3. Inlay or patch grafting.
- L** 4. Lattice or darning.
- 5. Repair in Layers – Cattle's technique.
- 6. Laparoscopic inlay mesh repair.
- K** 7. Keel operation.

Cattle's operation

Principle

The hernia is repaired in layers, which aims at anatomical restoration. The method is particularly suitable in small hernia where scarring is minimal.

Essentials of operation

After dealing with the sac and its contents the repair is done in multiple layers:

- *First layer:* Neck of the sack with an inter-locking suture of thick vicryl.
- *Second layer:* The cut edges of the base of the sac with unabsorbable sutures like prolene.
- *Third layer:* Matted tissue and fascia around the peritoneal sac with prolene.
- *Fourth layer:* Muscles are apposed with prolene.
- *Fifth layer:* Anterior rectus sheath with prolene.
- *Last layer:* Skin.

Keel operation

Principle

In this operation the hernial sac is not opened and the repair is done by wide inversion of the sac. In this repair the appearance of the inverted sac on cross section resembles the 'keel' of a ship (i.e., the bottom of the ship – Fig. 10.16); hence the name, Keel

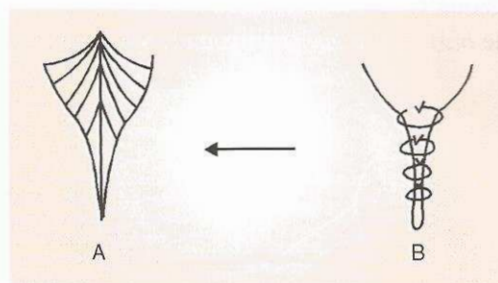


Fig. 10.16. Looking like the bottom of the ship (A); Keel operation (B).

operation (Rodney Maingot). So it is an *extra-peritoneal operation*. This method is particularly suitable for large ventral hernia.

Essentials of operation

After dissecting and cleaning the neck of the sac, the sac is pushed back into the abdominal cavity by a series of inverting and pleating layers of unabsorbable sutures (3 to 4 layers of sutures). Lastly, the anterior sheath and skin are repaired.

Advantages

1. Chance of peritonitis, postoperative ileus and abdominal distension is nil.
2. Early feeding may be started.

Disadvantages

1. As it is a blind operation (the sac is not opened) and the intra-abdominal pathology (e.g., any adhesions of the bowel and omentum with the sac) cannot be corrected, there may be a chance of subacute intestinal obstruction in late post-operative period.
2. During dissection of the sac and suturing of the sac after inversion there is a chance of injuring the bowel and omental vessels.

Lattice or darning

After dealing with the sac and its contents, the defect in the abdominal wall is closed with fascial sutures in an interlacing manner between muscle and aponeurosis of either side. *The interlacing suture is made in the form of lattice work or darning.*

Shoelace darning

There are two steps in this operation. *The first step is to reconstitute a strong midline new linea alba by suturing with prolene the strips of fasciae from the medial cut edges of each anterior rectus sheath which is incised longitudinally.*

The second step is to restore the recti muscles to their normal position beside the new linea alba without tension by drawing the lateral cut edges of the rectus sheath close to newly formed linea alba with prolene suture in a 'shoelace' darning fashion.

Inlay graft

These are becoming increasingly popular particularly

when the defect is very large and cannot be closed effectively by local tissues.

After dealing with the sac and its contents the peritoneum is sutured. Then *the defect in the abdominal wall is bridged across by implanting a sheet of tantalum gauze or dacron or marlex or polypropylene mesh*. The sheet of mesh should be tacked beneath the recti muscles and sutured all round with fine prolene sutures without tension—*inlay graft*. The recti muscles and anterior rectus sheath should then cover the mesh completely.

This repair may be supplemented with *onlay graft* which is placed on the outer aspect of musculo-aponeurotic abdominal wall deep to the skin and subcutaneous tissue – *combined inlay and onlay graft*.

Post-operative measures

1. Gastric suction to avoid distension.
2. Intravenous infusion of fluid and electrolytes till flatus is passed.
3. Avoid prolonged retention of urine and any other strain (e.g., cough).
4. Adequate antibiotic therapy.
5. Adequate nutrition and vitamins.
6. Early ambulation should be discouraged.
7. No strenuous work for three months at least.
8. Use of abdominal corset for 3 months.
9. Avoid weight gain and obesity strictly.

Incidence of recurrence after mesh repair

- Without mesh repair, incidence of recurrence is 30% - 40%.
- With mesh repair, the recurrence rate comes down to 10%.

Muscle pedicle flap

Recently many advocates the use of muscle pedicle flap to reconstruct the defect in the lower abdominal wall below the umbilicus. Two muscles are particularly suitable for this purpose – tensor fascia lata and rectus femoris.

Laparoscopic repair

Recently, laparoscopic repair of incisional hernia is being tried in some centres for small reducible hernia. After creation of pneumoperitoneum the adherent contents from the sac in the gap/gaps are released. Then the gap/gaps are covered intraperitoneally with

an appropriate size ($\geq 15 \times 15$ cm) mesh which is fixed to the abdominal wall with sutures and tackers.

UMBILICAL HERNIA

It is the herniation through or around the weak umbilical scar (cicatrix). It may be congenital or acquired and is of four different types:

1. Exomphalos.
2. Congenital umbilical hernia.
3. Infantile umbilical hernia.
4. Adult umbilical hernia.

EXOMPHALOS

Syn.: Omphalocele

It is a hernia occurring through the umbilicus into a transparent sac whose wall consists of peritoneum and amniotic membrane with intervening Wharton's jelly.

This rare hernia is due to failure of all or part of the midgut to return to the abdominal cavity during early part of intrauterine life (i.e., failure of second stage of gut rotation at about the beginning of the tenth week). At the same time there is failure of migration and fusion of the lateral folds of the anterior abdominal wall.

Pathology

The gut is found to be contained within a sac protruding through a defective umbilicus.

The *covering sac* when remains intact, looks like a thin semitranslucent membrane and consists of three layers — an outer layer of amniotic membrane, an intermediate layer of Wharton's jelly, and an inner layer of peritoneum (Fig. 10.16). The sac may occasionally be ruptured at delivery.

The *base of the sac* is surrounded by skin.

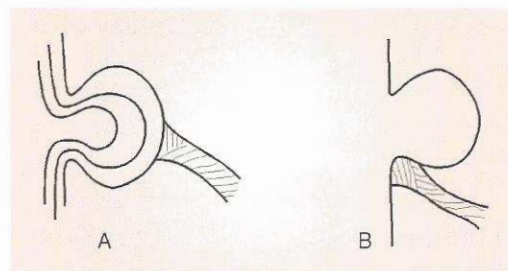


Fig. 10.17. Exomphalos: (a) Minor; (b) Major.

The *contents of the sac* vary. In the complete variety, the whole of the midgut loop occupies the hernial sac. In the partial variety, only the caecum and/or lower ileum occupy the sac.

Depending on the size of the fascial defect at the umbilical ring, exomphalos can be divided into minor and major.

Exomphalos minor

The size of the defect is less than 5 cm. The sac is relatively small in size, with the umbilical cord attached to its summit (Fig. 10.17A). The contents are usually one or two loops of bowel only. These loops may be inadvertently included in the ligature and cut across during severance of the cord.

Treatment

Twisting of the umbilical cord (which reduces the contents of the sac into the peritoneal cavity) followed by strapping of the abdominal wall at least for two weeks.

Exomphalos major

The size of the defect is more than 5 cm. The sac is larger in size, with the umbilical cord attached to its base near the inferior aspect (Figs. 10.17B & 10.18). The contents of the sac are always small and large intestines and occasionally a portion of liver.

On examination, the abdominal wall defect appears

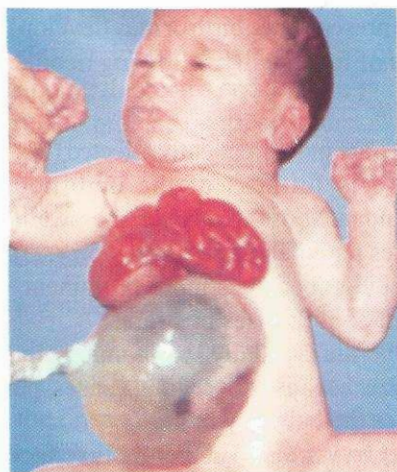


Fig. 10.18. Exomphalos major. The intestine prolapsed outside through a rupture of the sac at the upper part – just before taking the photo. (Courtesy: Dr. Abinash Roy, MD)

as a transparent veil through which the contents may be seen.

Majority of babies with exomphalos are reported to have associated anomalies such as:

- (a) Trisomy 13 or 18
- (b) Beckwith-Wiedmann syndrome
- (c) Cloacal exstrophy
- (d) Congenital heart disease.

Differential diagnosis

- Gastroschisis.

Treatment

- Conservative
- Operative

Conservative treatment

Indication:

1. In presence of associated other major congenital abnormalities.
2. In absence of facilities of operative repair, e.g., lack of experienced surgeon or well-equipped hospital.

Method:

Repeated application of 0.5 percent mercurochrome, or 0.25 percent silver nitrate and silver sulfadiazine cream over the sac is done followed by covering the sac with wet sterile gauge. This local application aims at promoting escharification of the sac which then shrinks and slowly gets epithelialised to form a ventral hernia. This hernia is repaired at a later date.

Operative treatment

Immediate surgical repair is preferable in presence of facilitates. Operation should be done within the first few hours of life, otherwise the sac will burst.

Different types of operations are available depending on the size of the defect, the general condition of the baby, and the presence or absence of other anomalies.

Pre-operative measures:

1. No feeding by mouth. Intravenous transfusion should be started.
2. Ryle's tube suction.
3. Blood transfusion should be arranged.
4. Antibiotics to be started.
5. The sac is protected with wet sterile dressings soaked in mild antiseptic or normal saline.

Types of operation:

- A. *Primary repair*: It consists of excision of the sac, reduction of the herniated viscera into the peritoneal cavity, and closure of abdomen in layers, all of which are performed in one stage.
- B. *Staged skin flap closure*: In the first stage, the umbilical cord is excised. The skin flaps are created by undermining the subcutaneous tissue on either side in the flanks. These flaps are then mobilised to facilitate closure over the intact sac. If necessary, lateral release incisions may be made and they are left to heal secondarily. Skin closure over the defect is performed in two layers. If the patient survives this operation, this leaves the child with a large ventral hernia. This ventral hernia is then repaired as a second stage procedure months or years later.
- C. *Staged silastic pouch closure*: In this procedure, a temporary closure of the abdominal wall is achieved by using Dacron reinforced silastic sheet in the form of a sac. At an interval of two days, the silastic sheet is shortened gradually, till it is finally removed when the approximated edges of the abdominal wall are sutured.

Whichever method of operation is adopted, the cardiopulmonary system must never be embarrassed by the tight closure of the abdominal wall. A tight closure may cause:

- (a) decrease in cardiac output due to decrease in venous return;
- (b) hypoxia following ventilatory insufficiency due to splinting of the diaphragm by the abdominal viscera.

The *survival rates* in exomphalos major are about 50 percent if there are no associated anomalies.

GASTROSCHISIS

1. In this condition there is a full-thickness defect in the abdominal wall either immediately adjacent to the umbilicus or separated from it by a thin strip of skin.
2. In gastroschisis, anomalies of other systems are rare. *But chances of obstruction are common.*
3. Gastroschisis needs urgent surgical intervention.
4. The survival rates in gastroschisis are better than exomphalos major, ranging up to 90 percent.

5. Operative treatment:

- (a) *Primary repair*: It consists of reduction of the herniated viscera into the peritoneal cavity and closure of abdomen in layers, all of which are performed in one stage.
- (b) Occasionally, *staged silastic pouch closure* may be required, as in exomphalos major.

Clinical differences between exomphalos and gastroschisis

Feature	Exomphalos	Gastroschisis
Defect	At the umbilical ring	Adjacent to the umbilical ring
Size of defect	Variable	Usually less than 5 cm
Sac	Usually present unless ruptured	Absent
Umbilical cord	Attached to sac	Normal in position
Herniated bowel	Usually normal	Oedematous, thickened and shortened
Herniation of liver	Common	Uncommon

CONGENITAL UMBILICAL HERNIA

It is a rare variety of well developed umbilical hernia occurring as a result of intra-uterine epithelialisation of an exomphalos minor.

INFANTILE UMBILICAL HERNIA

It is the herniation through the weak umbilical cicatrix. It may be present since birth or may develop in the first few days of life usually following neonatal sepsis.

Diagnostic criteria

1. Males are more frequently affected (M : F = 2 : 1).
2. There is fullness of the umbilicus with a redundant loose skin over the swelling – this causes parental distress and worry.
3. The bulge becomes more prominent when the infant or child cries or strains.
4. The hernia is usually symptomless, but increase in size because of crying causes pain which makes the baby cry more.
5. *On examination*: The hernia is reducible and a small gap can be palpated admitting the tip or tips of the fingers.
6. Obstruction or strangulation is very rare below the age of three years.

Treatment

Conservative

Most of the herniae close spontaneously without any treatment within two years of age. So, the methods are:

- Masterly inactivity.
- Reassurance to the parents.
- Customary strapping after reduction of the hernia: A coin covered with a pad is placed over the umbilicus, which is then fixed by adhesive strapping.

Operative

Herniorrhaphy is indicated when the hernia is still present after 2 years of age.

Essentials of operation:

1. The umbilicus should be preserved.
2. A small transverse curved incision is made immediately below the umbilicus with convexity towards the pubis.
3. The incision is deepened to expose the anterior rectus sheath.
4. After mobilisation of the skin flaps, the flaps with the umbilicus are retracted upwards.
5. The neck of the sac is identified and the sac is opened at the neck (cf. inguinal hernia where the sac is opened at the fundus).
6. Contents are reduced and the sac is ligatured and divided at the neck.
7. The defect in the linea alba is closed by overlapping the rectus sheath with two or three interrupted prolene mattress sutures.
8. The skin edges are accurately apposed and sutured.

ADULT UMBILICAL HERNIA

Syn.: Para-umbilical hernia

This is the common acquired umbilical hernia. It occurs not through the umbilical scar but through a defect in the linea alba just above or below or to the side of the umbilicus. *So, it is truly para-umbilical.*

Contributory factors

1. Commonly in middle aged or elderly women.
2. Obesity.

3. Multiparous women.
4. Persistent source of straining e.g., chronic cough, constipation, bladder neck obstruction, etc., leading to increased intraabdominal pressure.

Pathology

The hernia occurs through the weak area in the linea alba which is the result of repeated stretching and thinning out of the linea alba together with wide separation of the recti.

The sac is usually multiloculated. The neck of the sac is relatively narrower than the fundus because of its bulky contents. The superficial layer of the rectus sheath is stretched out over the protruding sac and is adherent to the sac. The overlying skin of the umbilical cicatrix may be adherent to the fundus of the sac in late cases.

Contents of the sac: Portions of small intestine, large intestine together with greater omentum. These contents are very often adherent to the fundus of the sac and also to one another.

Owing to the multiple loculi of the sac and to the presence of adhesions between the contents and the sac, the hernia is usually irreducible.

Diagnostic criteria

1. Common in the elderly, multiparous women, usually obese with a persistent source of



Fig. 10.19. Umbilical hernia through the umbilical cicatrix in 64 years aged male. No H/O constipation, urinary obstruction and asthma. The lump was reducible. Cough impulse positive. But the abdomen was distended with ascites. In an aged patient with umbilical hernia percussion for fluid thrill and shifting dullness should be performed to exclude ascitis. Management of ascites should take consideration first.

- straining like chronic bronchitis, chronic constipation and difficulty in passing urine.
- The patient presents with a swelling round about the umbilicus. She also complains of pain or discomfort and tenderness around the umbilicus. Such pain gets worse by prolonged standing or strenuous work. The patient may present with intermittent attacks of intestinal colic or subacute intestinal obstruction.
 - The swelling appears as a firm, round, knobby and pendulous mass.
 - Skin with the umbilical cicatrix hangs over the swelling like a festoon. The skin beneath the swelling may be unhealthy, infected or ulcerated.
 - Cough impulse is present in most cases.*
 - Most of the herniae are reducible at least partially.* They may be irreducible when the contents are adherent to the sac, or the neck of the sac is very narrow.
(The reducibility on lying down is not as early and easy as in incisional hernia because the neck of the sac in umbilical hernia is narrow whereas in incisional hernia it is wide.)
 - After reduction, a doughy mass due to adherent omentum may be felt.
 - On palpation*, the defect in the linea alba can be felt as firm fibrous edge, particularly in reducible hernia by insinuating the fingers along the margin of the swelling.
 - The recti may feel divaricated.



Fig. 10.20. Paraumbilical hernia. Swelling bulging through a defect in the linea alba below the umbilicus. Reducible. Cough impulse positive. No overlying scar.

Complications

- Irreducibility.
- Obstruction.
- Incarceration – more common than obstruction.
- Strangulation.

Treatment

Always operative because of the persistent risk of complications particularly when the hernia is irreducible. The standard practice is herniotomy followed by repair of the gap in the linea alba by *overlapping of the flaps of the anterior rectus sheath*, either from above and below (Mayo), or from side to side (Wells) and suturing them with interrupted stitches.

Pre-operative measures

- Correction of obesity by proper and controlled diet.
- Correction of any other contributory factors like chronic cough or constipation. Cardiac and renal functions are also to be investigated.
- Careful skin preparation: An important measure because the skin creases below the swelling are very often infected or ulcerated.

Mayo's operation

Principle of operation:

- The incision should be made transversely across the abdomen* to deal with the umbilical hernia.
Advantage: By the transverse incision, the fibres of the rectus sheath can be simply separated and so the abdominal wall is not further weakened. If a vertical incision is made, the fibres of the rectus sheath are also divided, thus adding to the already existent weakness of the abdominal wall (McGregor).
- After dealing with the hernial sac and its contents, the defective linea alba is repaired by overlapping of the flaps of the rectus sheath across a transverse axis (*Double-breasting technique*).
Advantages:
 - A satisfactory degree of overlapping is obtained without tension.
 - Tension to suture line is nil or minimum.

Essentials of operation:

1. A transverse elliptical incision is made encircling the umbilicus. The incision should extend laterally on either side beyond the swelling.
2. Incision is deepened to expose the aponeurotic sheath.
3. Mobilisation of upper and lower skin flaps to expose aponeurotic sheath adequately *for about 3 inches, both above and below, beyond the rim of the gap*. Edge of the defect is well-defined.
4. The neck of the sac is identified and cleaned first.
5. *The neck of the sac should be opened first* because the contents are adherent to the fundus, and the neck remains free from adhesions (cf. Inguinal hernia).
6. Dealing with the contents of the sac: Any adherent omentum and bowel are carefully dissected, freed and returned to the abdominal cavity. Small portion of sac may be left attached with the gut. Bulky adherent omentum may be excised taking care of the colon.
7. The redundant sac along with the overlying skin is excised, and the peritoneum of the neck is closed with catgut.
8. Creation of upper and lower flaps of the anterior rectus sheath by elongating the transverse incision on either side of the defect sufficiently to allow adequate overlapping of the flaps. The flaps should be cleared of any fatty tissue.
9. Repair of the defect by overlapping of the flaps including the peritoneum using four or five non-absorbable mattress sutures. *Usually the lower flap is placed underneath the upper flap.*
Each suture should pass through all the layers of the flaps.
The margin of the upper flap is next sutured with the lower flap by a continuous or interrupted stitches.
10. Skin is closed with a subcutaneous drain at each end of the wound, preferably a vacuum drain, which should be kept for 48 to 72 hours.

Post-operative measures

As discussed under incisional hernia.

EPIGASTRIC HERNIA

Syn.: Pre-peritoneal lipoma; Epigastric lipoma; Fatty hernia of linea alba

It is the protrusion or herniation of extra-peritoneal fat through a defect in the linea alba anywhere between the xiphoid process and the umbilicus, usually midway between these structures.

Often it is called 'epigastric lipoma' though it is a misnomer.

Contributing factors

1. Always acquired.
2. More common in manual labourers between the ages of 30 and 45 years.
3. Sudden strain causes tearing of the interlacing fibres of the linea alba.

Pathological event

In the first stage there is protrusion of extra-peritoneal fat through a defect in the linea alba, usually where the latter is pierced by a small blood vessel. At this stage there is no well-formed sac but only the extruded extra-peritoneal fat, and so it is known as *pre-peritoneal lipoma*. This lipoma is attached by a pedicle to the underlying peritoneum.

In the next stage this extruded extraperitoneal fat or lipoma grows bigger and bigger to come out of the defect and drags a pouch of peritoneum after it and thus gives rise to a true epigastric hernia. *The lipoma is attached to the apex of the peritoneal sac.* The mouth of the sac is very narrow and does not permit a part of hollow viscus to enter into it. The sac is usually empty or it may contain a small portion of the greater omentum.

Diagnostic criteria

1. Subject: Common in manual labourers.
2. Age: 30 to 45 years.
3. Presenting symptoms: May be:
 - *Symptomless*: No symptoms. Discovered only during routine abdominal palpation.
 - *Pain*: The patient complains of epigastric pain which is localised exactly to the site of the hernia and worsens on physical exertion but often he does not notice any lump.
 - *Lump in the epigastrium*: Painless, or painful when the lump may be tender to touch or tight clothing. (It is possibly because the fatty contents become nipped sufficiently to produce partial strangulation.)
 - *Referred pain*: Often the patient does not notice the epigastric lump. But he complains

of epigastric pain suggestive of a peptic ulcer or gall bladder disease. In many cases of epigastric hernia there is co-existent upper abdominal disease.

4. *On examination:*

- A firm swelling in the epigastrium.
- Does not usually have cough impulse.
- Cannot be reduced.
- May be tender.

Treatment

If small and no symptoms – the lump can be overlooked.

If there are symptoms – operation. But before undergoing any operation the patient should be thoroughly investigated to exclude any intra-abdominal pathology, e.g., peptic ulcer or disease of gall bladder.

LUMBAR HERNIA

Two types:

1. Primary
2. Secondary:
 - Following an operation upon an infected kidney or drainage of lumbar abscess, in which the wound gets infected post-operatively – the example of incisional hernia. So, an incisional scar is noted in and around the swelling.
 - Following local paralysis of the anterolateral abdominal muscles, e.g., due to poliomyelitis – also known as Phantom hernia.



Fig. 10.21. Right-sided lumbar hernia – cough impulse positive and reducible.

PRIMARY LUMBAR HERNIA

The hernia protrudes through the lumbar triangle of Petit in the posterior abdominal wall.

Boundary of Petit's triangle

- *Base* – formed by the crest of the ilium.
- *Anterior border* – formed by the posterior free margin of external oblique muscle.
- *Posterior border* – formed by the anterior margin of latissimus dorsi muscle.
- *Floor* – formed by the internal oblique and transversus abdominis muscle. The floor must be weak or absent to allow the hernia to occur.

The hernial sac has usually a wide neck. The sac may contain right or left colon.

Diagnostic criteria

1. Swelling in the lumbar region just above the iliac crest.
2. Soft swelling.
3. Cough impulse – present.
4. Reducible – on lying down or on pressure.

Differential diagnosis

1. Lipoma: Firm to solid swelling. Cough impulse absent. Not reducible.
2. Para-vertebral cold abscess pointing to this position – soft cystic swelling, fluctuation positive. Cough impulse absent. Not reducible. Both clinical and radiological examinations of the spine will show evidence of tuberculosis of vertebrae.
3. Incisional lumbar hernia: Previous history of operation followed by wound infection and local scar are present.

Treatment

Operation: Herniotomy + Herniorrhaphy, or Hernioplasty with a piece of tentalum gauze or dacron mesh.

SPIGELIAN HERNIA (Fig. 10.22)

This hernia protrudes through the linea semilunaris along the lateral border of the rectus muscle at the level of the arcuate line. It appears as a small, tender swelling to one side of the lower abdominal wall along the lateral margin of the rectus muscle. Operation is the treatment of choice.

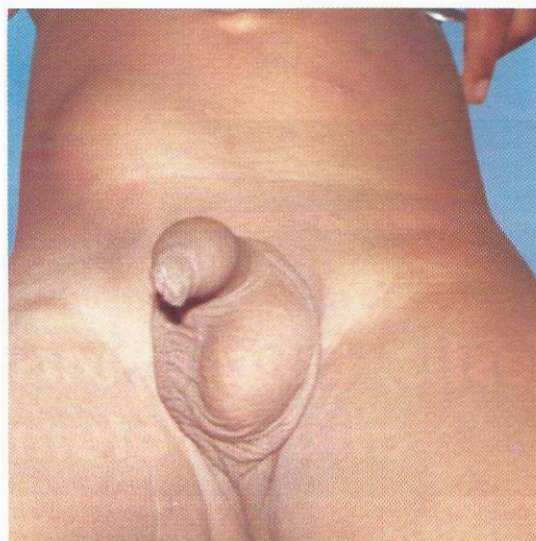


Fig. 10.22. Right spigelian hernia + right-sided undescended testis. Abdominal swelling was reducible and cough impulse was positive – so, a hernia. Same patient had right-sided undescended testis. On exploration, hernial sac was found protruding through the lateral margin of rectus abdominis. The content was found to be right testis – a surprising content of the hernia at this site. Testis was mobilised and implanted to the right scrotum. Hernia was repaired.

Differential diagnosis of a lump in the groin

When a patient presents with a lump in the groin one must think of anatomical structures in and around the area and correlate the pathological lesions arising from the structures.

1. The hernial orifices

- Inguinal hernia
- Femoral hernia

2. *The lymph nodes*
 - Lymphadenopathy
 - Inflammatory
 - Neoplastic
 - Reticuloses
3. *The vein*
 - Saphena varix
4. *The artery*
 - Femoral aneurysm
5. *The testicular apparatus*
 - Hydrocele of the cord or canal of Nuck
 - Lipoma of the cord
 - Undescended testis
 - Ectopic testis
6. *The psoas sheath*
 - Psoas abscess
7. *The skin and subcutaneous tissues*
 - Lipoma
 - Neurofibroma
 - Sebaceous cyst.

OBTURATOR HERNIA

This rare hernia protrudes through the obturator foramen and proceeds into the medial side of the thigh. Irreducibility, intestinal obstruction and strangulation are more common with this hernia. Many a time patient presents with intestinal obstruction when exploratory laparotomy only can settle the diagnosis of obturator hernia as the cause of intestinal obstruction.

11

CHAPTER



Testis, Epididymis and Scrotum

Disorders of the testis and epididymis are commonly encountered in clinical practice. Most of them give rise to solid or cystic scrotal swellings with the exception of imperfect testicular descent.

Classification of disorders

Testis and epididymis

1. Imperfect descent
2. Inflammation
3. Tumours
4. Cysts
 - Cysts connected with the epididymis
 - (a) Cyst of the epididymis
 - (b) Spermatocoele.

Spermatic cord

1. Torsion
2. Varicocele

Tunica vaginalis

1. Hydrocele
2. Haematocele.

CRYPTORCHIDISM

Cryptorchidism means a hidden testis. When absent from the scrotum, the testis is hidden from the sight.

Before we consider the pathology under this heading we should discuss the development of the testis.

Development of testis

The testes develop retroperitoneally in the lumbar region from the genital ridge on the medial aspect of the mesonephros (Wolffian body), and so in early life they lie in the coelomic cavity.

The primitive testis is attached to the posterior abdominal wall by a fold of peritoneum, the mesorchium, which contains the testicular blood vessels, and nerves derived from the tenth and twelfth dorsal segments respectively (hence the testicular pain is referred to the umbilicus). Most of the Wolffian body disappears, but the Wolffian duct persists as the epididymis and vas deferens.

A strand of fibromuscular mesoblastic tissue (the gubernaculum) is attached to the lower pole of the testis and bottom of the scrotum. It acts as a rudder to guide the descent of the testis to its normal position, i.e., scrotum (Gubernaculum means rudder).

During its descent the testis is preceded by a fold of peritoneum (processus vaginalis) which projects into the foetal scrotum.

Contraction of the gubernaculum, together with increased intra-abdominal pressure, brings about the normal descent of the testis.

Timetable of testicular descent (Fig. 11.1)

- Third month of foetal life – iliac fossa.
- Seventh month of foetal life – deep inguinal ring.

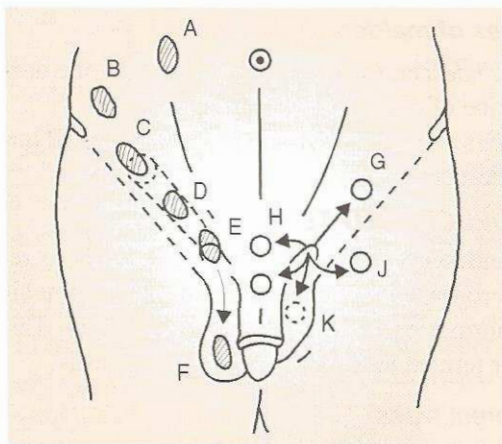


Fig. 11.1. The time table of testicular descent on the right side (these are also the different positions of undescended testes). A, lumbar region; B, iliac fossa; C, deep inguinal ring; D, inguinal canal; E, superficial inguinal ring; F, scrotum. On the left side showing common positions of ectopic testes: G, superficial inguinal pouch; H, pubic; I, penile; J, femoral triangle; K, perineum.

- Later part of the seventh month of foetal life – travelling down the inguinal canal.
- Eighth month of foetal life – superficial inguinal ring.
- By nine months i.e. shortly before or at birth – drops into the scrotum, i.e., the final seat of destination.
- Right testis descends little later than the left testis.

Factors helping the descent of testis

1. Shortening of and traction of the gubernaculum testis.
2. Differential growth of the body wall in relation to a relatively immobile gubernaculum.
3. Raised intra-abdominal pressure due to foecal accumulation and relative growth of the abdominal viscera – pushing the testis through the inguinal canal that is engorged by the swollen gubernaculum.
4. Development and maturation of the epididymis inducing testicular descent.
5. Higher temperature inside the abdomen, which is detrimental to spermatogenesis.
6. Hormonal factors – supposed to play the major role in promoting testicular descent. The hormones are chorionic gonadotrophin from maternal circulation, testosterone and dihydrotestosterone from the testes.

Factors interfering with the descent of testis

1. Retroperitoneal adhesions.
2. Obstruction e.g., lateral adhesion at the deep inguinal ring.
3. Short vas deferens.
4. Short testicular vessels.
5. Short pampiniform plexus.
6. Inefficient pull by the gubernaculum testis.
7. Deficient hormonal stimulation.
8. Imperfectly developed testis (e.g., dysgenesis) which makes the testis insensitive to gonadotrophins.
9. Deficiency of gonadotrophin and/or testosterone.
10. Deficiency of Mullerian inhibiting substance which is secreted by fetal Sertoli cells which is essential for testicular descent.
11. *Prune-Belly syndrome* leading to poorly developed abdominal wall.
12. Maternal exposure to exogenous estrogens (oral contraceptives) during the first trimester of pregnancy has been identified as a risk factor.

Pathological changes of undescended testis

1. *Growth of the testis:* An undescended testis fails to develop normally. After the age of six years growth of the testis is retarded and the testis becomes hypoplastic. By the time of puberty the incompletely descended testis is soft and flabby and hardly more than half the size of its intra-scrotal counterpart. Testis becomes soft and flabby due to atrophy in view of the fact that scrotum acts as a thermoregulator to the testis maintaining the temperature at least 2 °F lower than that of the inguinal canal, but the undescended testis is subjected to a higher temperature.
2. Epididymis is separated from the testis by a long mesorchium – a factor for torsion of undescended testis.
3. An incompletely descended testis is often associated with a hernial sac.
4. *Histological changes:* Epithelial elements are grossly immature. By the age of sixteen irreversible, destructive changes occur in the germinal epithelium. Interstitial cells development will however proceed normally.

5. *External secretory function:* The spermatogonia content starts diminishing from the age of 2 years, and spermatogenesis becomes feeble over the months or years and ultimately ceases; unless the testis is placed in the scrotum by the age of ten years spermatogenesis will be defective. When the condition is unilateral, the spermatogenesis is carried out by the normally descended testis. Bilateral cases invariably develop sterility.
6. *Internal secretory function:* Interstitial cells (Leydig cells) development will, however, proceed normally and *secondary sex characters* appear. In bilateral cryptorchism about half the normal amount of androgen is produced. Nevertheless, the secondary sex character is noticeable in most of the cases and endocrinologic cause of impotence is rare.
7. Post-natal growth of testis passes through three stages:
 - The resting stage (0–4 years).
 - The growth stage (4–10 years).
 - The maturation stage (10 years and throughout adolescence).

At the growth stage spermatogenesis begins to appear and seminiferous tubules become tortuous and it is at this stage that an abnormally placed testis undergoes imperfect but reversible spermatogenesis; permanent and irreversible changes at the maturation stage will occur if the testis is not in the scrotum by the 10th year.
8. Cryptorchism may be associated with abnormalities of urinary tract system (13.3%).

Incidence

- Approximately 4 percent of all full-term newborns have an undescended testis or testes (about 80% cases are unilateral). It may be bilateral (20%).
- The frequency is significantly higher in premature neonates (20%).
- By one year of age, 75% of full terms and 95% of premature cryptorchid testis descend spontaneously.
- Family history is positive for cryptorchidism in roughly 15% cases.
- Undescended testis is more common in the right side (50 percent) than in the left side (30 percent).
- *Why common in the right side in unilateral cases?* Because the right testis usually descends later than the left testis.

Types of maldescended testis

- A. *Undescended testis:* Arrested along the normal line of descent.
- B. *Ectopic testis:* Deviated from the normal line of descent.

UNDESCENDED TESTIS

An undescended testis is one which fails to reach the scrotum but which is retained at any point along the normal path of its descent (Fig. 11.1). This is better termed incompletely descended testis.

Different types

Based on the location of the testis (can be remembered by the mnemonics 'AIRES').

- A. **Abdominal:** Retroperitoneally in the posterior abdominal wall.
- I. **Iliac:** Deep to the deep inguinal ring.

Inguinal or interstitial: Lies in the inguinal canal.

In early life, the testis is soft and lies over the non-resisting floor of the inguinal canal and is covered by the overlying tendinous aponeurosis of the external oblique; so this variety is also clinically difficult to palpate.

- R. **Retractile testis:** Here, due to contraction of over-active cremaster muscle, the testis is pulled upward either in the inguinal canal or in the superficial inguinal pouch (a space lies superficial to the aponeurosis of external oblique but deep to the fascia of Scarpa. The space lies lateral to the superficial inguinal ring).

So, in retractile variety the testis can be coaxed down in the scrotum i.e., its normal position. The scrotal sac is also normal. The testis is usually of normal size. This variety of testis requires neither endocrine nor operative treatment. In course of time (may be delayed up to puberty) the testis will take up its normal position permanently.

- E. **Emergent testis:** Movement of the testis ranges from the inguinal canal to the superficial ring. Emergent testis is transiently palpable outside the superficial inguinal ring, but liable to slip back again into the inguinal canal.
- S. **Simple undescended or high retractile testis:** Movement of the testis ranges from the superficial inguinal ring to the upper scrotum.

ECTOPIC TESTIS

An ectopic testis is one which fails to descend into the scrotum but has also deviated from its usual path of descent.

Ectopic testes are far less common than incompletely descended testis.

An ectopic testis may be found in one of the following positions, in order of frequency (Fig. 11.1).

1. *Superficial inguinal pouch* (i.e., above and lateral to the superficial inguinal ring and superficial to the external oblique aponeurosis) – commonest site.
2. *Suprapubic* i.e., at the root of the penis.
3. *Perineum*.
4. *Femoral triangle*.
5. *Opposite scrotal compartment* – transverse testicular ectopia is the rarest of all ectopic testes.

Explanation regarding ectopic position of testis

According to Lockwood, the gubernacular band has got 4 other additional or accessory tails known as tails of Lockwood, in addition to the scrotal tail. These are (Fig. 11.2):

1. Superficial inguinal tail
2. Pubic tail
3. Perineal tail
4. Femoral tail.

Normally all these accessory tails disappear, not the scrotal tail. But in ectopic testis the scrotal tail becomes ruptured. As a consequence, the testis follows one of the accessory rudders, and takes up the position as mentioned above.

Diagnostic criteria (cryptorchidism)

Symptoms

1. Parents seek advice for the baby because of empty scrotum either one side or both sides due to absence of testis.

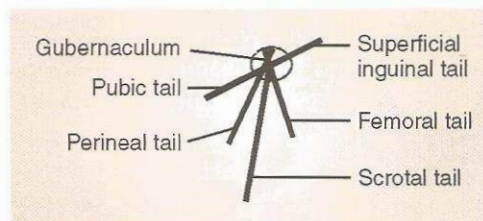


Fig. 11.2. 'Gubernacular tails' of Lockwood.

2. If the parents fail to notice, the patient may notice it during adolescence or in adult life.
3. A small proportion of patients may present in adult life because of infertility.

Local examination

4. The scrotum is not developed in undescended side and so looks asymmetrical. When both testes are undescended it is small and hypoplastic.

In retractile and ectopic varieties the scrotum is normally developed.

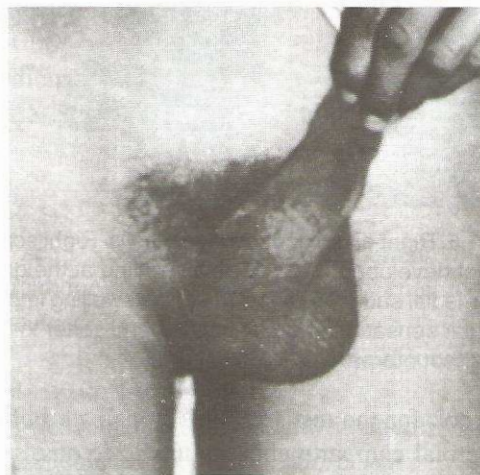


Fig. 11.3. Right-sided undescended testis. Right testis was not palpable in the groin. Right scrotum was not developed.

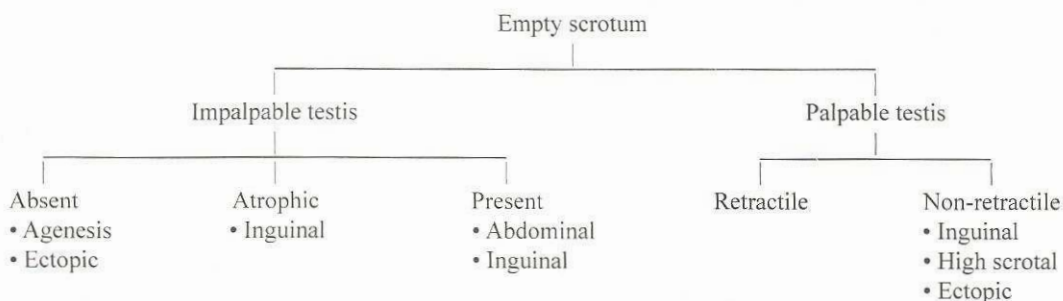




Fig. 11.4. Right-sided undescended testis. Right scrotum was not developed (cf. retractile and ectopic testis). Look for the testis – testis is not palpable anywhere in the groin.



Fig. 11.6. Bilateral undescended testis. Lump noted on the right groin suggestive of testis. Left testis was not palpable.



Fig. 11.5. Right-sided undescended testis. Right scrotum was not developed. There was a swelling at the groin – at the medial end of inguinal canal. The swelling imparted testicular sensation and was smaller and softer than the normal counterpart.

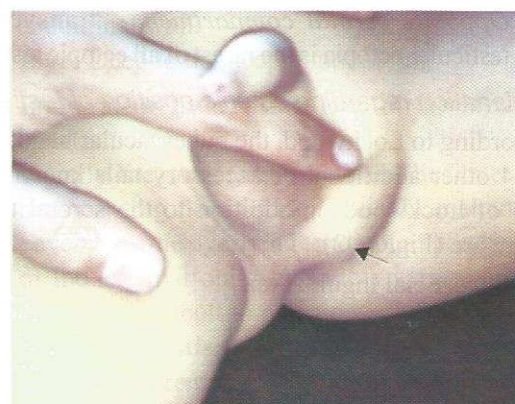


Fig. 11.7. Left ectopic testis in the perineum. Right scrotum developed with right testis. Left scrotum not developed. Left testis noted in left perineum (marked by arrow). (Courtesy: Prof. Sugato Banerjee, MS, MCh)

5. Look for the testis. Carefully palpate both the scrotal compartments, inguinal regions, suprapubic area, perineum and femoral canal. The testis may be impalpable or palpable.

Impalpable testis

It may lie in the abdomen or inguinal canal; it may be absent or atrophic.

- An abdominal or inguinal testis may become palpable on coughing when accompanied by a hernia.
- The undescended testis in the inguinal canal is very difficult to feel but every attempt should be made to coax it down to the superficial inguinal ring by pushing it medially and downwards so that the external oblique aponeurosis

no longer covers it, thus rendering it palpable. *Pressure along the line of the inguinal canal from the lateral to medial end may evoke tenderness or testicular sensation.*

Palpable testis

It may be either inguinal, high scrotal, retractile, or ectopic.

- *Retractile testis:* The testis can be coaxed down to the scrotum.
- *High retractile testis:* The testis is usually palpable just beyond the superficial inguinal ring but cannot be coaxed down into the scrotum.
- *Non-retractile palpable testis:* The testis is either high scrotal, inguinal or ectopic.

6. The testis is smaller in size and soft in consistence in undescended variety. So the undescended testis is very difficult to feel.
In ectopic and retractile variety the testis is normal sized and lying very superficially.
7. Testicular sensation: Indicates that the swelling is testis. In case of a child, while the testis is pressed the child will cry.
8. Range of mobility should be looked for to differentiate the different varieties. It also indicates the planning of operation.
9. Look for the presence of any complication – particularly hernia. Undescended testis almost always will have an associated hernial sac. So, look for a cough/cry impulse in the inguinal region.
10. Patients with unilateral undescended testis will have a normal puberty and secondary sex character. Patients with bilateral undescended testes are inclined to be fat with poorly developed sexual character. In cases of bilateral undescended testes, if the penis is very small and/or is associated with hypospadias, one should suspect an “intersex” problem.
(One should be cautious about the overweight boys who always appear to have small genitals, as the penis is swallowed up in the abdominal fat and the scrotum appears smaller than what it should be; parents should be reassured about this and advised to take measures to reduce the body weight.)
11. Look for the other congenital abnormalities.

How to differentiate between the undescended testis in the inguinal canal and the ectopic testis in the superficial inguinal pouch?

There are two valuable physical signs:

1. The patient is asked to tense his abdominal muscles (and hence the external oblique aponeurosis) by raising his head and shoulders from the couch – in case of ectopic variety the testis becomes more prominent and easier to feel as it lies on the taut external oblique aponeurosis. Moreover the mobility of the testis will be unaffected.
In undescended variety the testis becomes less prominent or impalpable.
2. The ectopic testis, when pushed laterally and upwards, becomes more prominent and goes merely to a higher level, superficial to the aponeurosis.
The undescended testis within the inguinal canal disappears through the deep inguinal ring.

How to differentiate between retractile testis in the superficial inguinal pouch and ectopic testis in the superficial inguinal pouch?

In retractile variety, the testis can be coaxed down into the scrotum but not in case of ectopic variety.

Investigations for undescended testis

Though investigation plays a minor role in the diagnosis of an empty scrotum, the following may be helpful in locating the testis particularly the intra-abdominal testis.

- Selective gonadal venography for proving the presence and position of a testis. Demonstration of a pampiniform plexus makes it almost certain that a testis is attached.
- Herniography.
- Radionuclide testicular scan.
- Ultrasound scan – Testis within the inguinal canal or positioned just inside the internal ring can be detected. Intraabdominal testis is difficult to detect with US.
- CT scan – Useful in prepubertal patient when the intraabdominal testis is sufficiently enlarged to be detected.
- MRI – Shows high success rate in determining the non-palpable testis.
- Laparoscopy – Most useful in identifying the impalpable testis or testes: *anorchism or agenesis. It has become the first choice of investigation recently. Moreover, therapeutic measure can be taken at the same sitting.*
- Hormone studies: Serum testosterone, LH, FSH, in case of bilateral undescended testis.

Complications of maldescended testis

Can be remembered by the mnemonics ‘SATHI’

- S** Sterility
- A** Atrophy
- T** Trauma/Torsion/Tumour
- H** Hernia
- I** Inflammation

Sterility or infertility

It is the rule in bilateral cryptorchid testes, treated or untreated. The fertility of patients with unilateral cryptorchid is also poor because of the usual defect in spermatogenic activity of the contralateral scrotal testis also. Evidence of decreased fertility exists even in unilateral cryptorchid patients who had undergone prepubertal orchidopexy. The prospects of fertility (spermatozoa count > 20 million/ml and mobility > 30%) after unilateral orchidopexy may be as high as 80 percent).

Atrophy of the testis

It occurs if descent does not occur by puberty. It may occur even earlier when the testis is situated in the inguinal canal due to repeated trauma.

Very rarely, atrophy rather than a lump is the first sign of malignancy, and so may be an indication for orchidectomy supplemented with prosthetic implantation.

Occasionally, the patient may present with the complaint that a previously small atrophic testis has enlarged to the size of the normal contralateral testis – it should arouse *suspicion of malignancy in atrophic testis*. Malignancy can develop also in atrophic testis.

Trauma

An inguinal testis is liable to oft-repeated trauma, giving rise to pain and atrophy.

Torsion

Because the undescended testis is very often suspended by a narrow mesentery and also because of the abnormal anchorage of the testis to the scrotum. Torsion of an imperfectly descended testis gives rise to painful tender swelling which is difficult to differentiate from a strangulated hernia or epididymo-orchitis.

Tumour

The risk of malignant change in an undescended testis is 50 times greater than in a fully descended testis. Approximately 10% of all testicular tumours occur in an undescended testis. Chances of malignancy are:

- higher in abdominal type (1 in 20), and
- lower in inguinal type (1 in 80).

In case of unilateral undescended testis, the opposite palpable testis which has normally

descended on other side, is also more prone to malignant change than normal individual.

All types of malignant testicular tumour may develop in undescended testis, *but seminoma is commonest followed by embryonal carcinoma*.

Orchidopexy does not reduce the risk of malignancy, but facilitates easy and early detection.

The appearance of a lump in the line of testicular descent, whether within the abdomen or the inguinal canal, and an empty scrotum should arouse the suspicion of malignant change in the undescended testis.

Hernia

Either indirect inguinal hernia or interstitial hernia is often associated with the undescended testis (about 90%).

Inflammation

Epididymo-orchitis is rare because the abnormal testis is less likely to become involved by descending infection.

Right-sided epididymo-orchitis may simulate appendicitis.

Treatment**1. UNDESCENDED TESTIS**

Surgery is the treatment of choice. Hormonal therapy has no role in this case.

Procedures available

1. Orchidopexy, i.e., fixation of the testis in the scrotum.
2. Orchidectomy, i.e., removal of the testis.

Orchidopexy**Idea of orchidopexy**

1. To allay worries and anxiety of both parents and patients – to improve cosmetic and psychological outcome.
2. To maximise the prospects of fertility.
3. To minimise the risk of tumour development.

Though orchidopexy does not confer immunity against malignancy, it may help screening for malignancy easier by placing the testis in a readily palpable position.

4. To reduce the risk of torsion.
5. To repair the associated inguinal hernia.

Principles of operation

1. Mobilisation of testis and the spermatic cord to obtain adequate length of the spermatic vessels allowing placement of the testis into the scrotum.
2. Repair of any associated hernia.
3. Fixation of the testis in the scrotum (orchidopexy), preferably without tension.

Ideal time for operation

All testicles should be brought into the scrotum by the age of 5 years to improve the spermatogenesis power. Postponement of operation beyond the age of 5 years results in germ cell atrophy. Moreover, early orchidopexy also reduces the incidence of malignancy.

Prior to 3 years of age, the cord structures are so small that operation may endanger the arterial supply to the testis.

So, ideally, orchidopexy should be performed between 3 and 5 years of age i.e. after the child is dry, before he goes to school and before any permanent histological changes have developed.

In bilateral cryptorchidism, one side is operated upon at a time with an interval of 6 months between the operations.

Methods

Various methods of fixing the testis in the scrotum are available and each merits its advantage according to the length of the cord after mobilisation.

1. Ladd & Gross technique.
2. Ombredanne's technique.
3. Keetley-Torek technique.
4. Placing the testis in 'Dartos pouch'.

Ladd & Gross technique

In this technique the testis is placed into the scrotal sac followed by narrowing of the neck of the scrotum. The testis is anchored below by passing a non-absorbable stitch through the tunica albuginea at the lower pole of the testis, and the stitch is brought out through the bottom of the scrotum, and anchored to the skin of the thigh by tying it over a piece of rubber tube. This tension apparatus is left in place for 10 to 12 days.

Disadvantage: Chance of recurrence after removal of tension stitch.

Ombredanne's technique (Fig. 11.8)

In this technique the testis is placed in the contralateral scrotal compartment by traversing the median septum of the scrotum. After repositioning the testis, the hole in the septum is narrowed taking care so as not to make it too narrow to cause obstruction of the structures of the cord.

The method is difficult to perform, and has no special advantage.

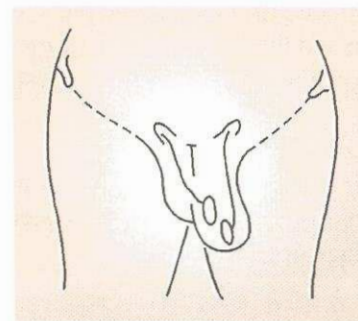


Fig. 11.8. Ombredanne's technique.

Keetley-Torek technique (Fig. 11.9)

In this technique the testis is placed into a pocket made into the upper and inner aspects of the thigh superficial to the fascia lata, adjacent to the scrotal sac. The testis is brought out through a scrotal incision and either the tunica albuginea covering its lower pole or the stump of the gubernaculum is stitched to the fascia lata of the thigh. Margins of the scrotal incision and thigh incisions are then united. This tension apparatus is maintained for 3 to 6 months.

At a second operation, 3 to 6 months later, the scrotum and the testis are separated from the thigh and thus the testis is replaced into the scrotum.

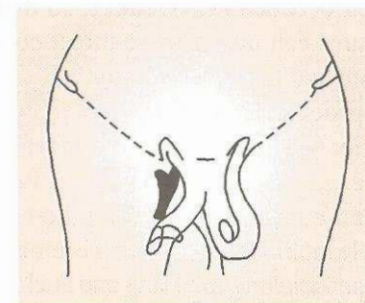


Fig. 11.9. Keetley-Torek technique.

Disadvantages:

1. The child has to undergo two operations.
2. There is considerable tension on the testis which is likely to interfere with the blood supply hence may lead to atrophy.

Placing the testis in the dartos pouch (Fig. 11.10)

After mobilising the cord and testis, the testis is placed in the scrotum. To keep it in the scrotum, a space is formed artificially in the plane between the scrotal skin and the dartos muscle (*dartos pouch*) at the bottom of the scrotal sac and the testis is placed there.

Advantages:

1. Rubber bands and traction are not necessary, if the mobilisation of the cord has been adequate.
2. One stage operation.

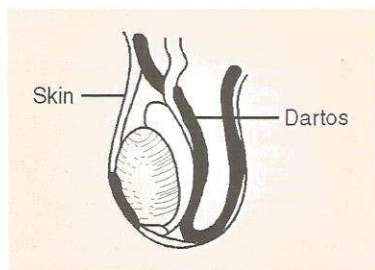


Fig. 11.10. Placing the testis in dartos pouch.

Problems during operations

Inability to obtain sufficient length of the cord to bring the testis down. Following methods are available to deal with this problem:

1. During mobilisation, if necessary, the fascia transversalis may be incised medially from the deep ring, and the inferior epigastric vessels are divided between the ligatures, so that the cord structures can take a more direct course to the scrotum and thus gain in length. This procedure would seem to weaken the posterior wall of the canal, but tendency to hernia is little.
2. *Two-stage orchidopexy:* In the first stage, after possible mobilisation the testis is brought outside the superficial inguinal ring and anchored as low down as possible by non-absorbable suture. At a second stage, after about months to 1 year a tedious re-exploration and attempt to obtain

sufficient length can be made followed by orchidopexy.

3. *Fowler-Stephens procedure:* The technique relies on the assumption that the collateral blood supply, primarily from the anastomoses between the artery to the vas and the testicular artery, will prevent infarction of the testis. The testicular vessels are clamped atraumatically for 5 minutes, and then the tunica albuginea is incised. If there is brisk bleeding from the testis it is assumed that the collateral blood supply is sufficient to obtain a viable testis. The testicular vessels are then sacrificed and the testis is placed into the scrotum.
4. *Silber procedure of microvascular anastomosis of the testicular vessels:* With the aid of operating microscope the testicular artery is anastomosed to the inferior epigastric artery and the testicular vein to the epigastric vein, but the vas and vasal artery are left undisturbed.
5. *Orchidectomy, in unilateral cases.* It may be followed by either simultaneous or subsequent placement of a testicular prosthesis to avoid emotional trauma.

Orchidectomy (removal of the testis)

Indication

1. The undescended testis is either hopelessly atrophic or cannot be brought down due to insufficient length of the cord structures, but the contralateral testis is normal.
2. Unilateral undescended testis in adolescent or adult, because of the risk of malignancy.
3. Bilateral undescended testes — when neither testis can be brought down, the smaller testis is removed and the larger one is preserved for its androgenic function.

If the testis cannot be found in the inguinal region it is advisable to look for an abdominal testis and remove it because of the risk of malignancy.

Laparoscopic management of undescended testis/cryptorchidism

- Recently laparoscopic intervention is in favour of the treatment of undescended testis (anorchism).
- Laparoscopy has both diagnostic and therapeutic roles to play in the management of undescended testis in the same sitting.

- Intraabdominal testis lying proximal to internal inguinal ring is difficult to be brought down to scrotum by conventional/open inguinal approach.
- Laparoscopy helps to detect the location of the undescended testis: *peeping testis*, *testis lying at the internal inguinal ring* or *testis lying 2.5 cm proximal to the internal ring or above*.
- Laparoscopy offers the choices of single stage as well as two-staged orchidopexy.
- Laparoscopy offers to treat bilateral undescended testes in the same sitting.
- Laparoscopy may detect the absent (agenesis) or atrophic testis.
- Testicular absence (agenesis) can be established *only when blind ending testicular vessels are found*.
- Atrophic testis should be removed when contralateral testis is in normal position, because tumour may also develop in atrophic testis.
- The laparoscopic orchiectomy is a simple procedure in experienced hand.
- *First diagnostic laparoscopy is performed before hormonal studies*: the absence of bilateral testicular vessels or blind ended testicular vessels suggest that the testes are absent on both sides—*agenesis*.
- Then patient is referred to endocrinologist for hormonal studies: serum Testosterone, LH and FSH followed by hormonal stimulation test.
- Serum testosterone level is normal in unilateral undescended testis as the other testis is normal.
- Serum testosterone level is also normal in bilateral undescended testis, provided they are not a case of bilateral agenesis.
- Hormonal stimulation study is available to distinguish bilateral agenesis from bilateral undescended testes.
- A short course of human chorionic gonadotrophin (hCG-2000 IU daily for 3 days) is given. In bilateral agenesis basal serum gonadotrophin (FSH and LH) levels are extremely high with a low serum testosterone level—the low serum testosterone level does not respond to the challenge with hCG.

Diagnostic laparoscopy and Fowler-Stephens procedure

Fowler-Stephens procedure can be performed laparoscopically also in two-stage procedure or one-stage procedure for high intra-abdominal testis (> 2.5 proximal to internal ring).

- *Two-stage procedure*: In the first stage the testicular vessels are clipped in situ with a hulk clip. In the second stage, six months later, the testicle is brought down after division of the testicular vessels into the dartos pouch of the scrotum based on the vasal vasculature.
- *One-stage procedure*: The testicular vessels are clamped atraumatically for 5 minutes, and then the tunica albuginea is incised. If there is brisk bleeding from the testis it is assumed that the collateral blood supply is sufficient to obtain a viable testis. The testicular vessels are then sacrificed and the testis is brought down in the dartos pouch of the scrotum.

Problem with impalpable testis or testes—Whether cryptorchidism or agenesis

When bilateral:

The diagnosis of bilateral agenesis or anorchism should always be considered in the presence of bilateral impalpable testes.

When unilateral:

- Unilateral agenesis poses a unique diagnostic problem. Hormonal studies and stimulation test does not help.
- Testicular venography may be useful: The absence of pampiniform plexus or a blind ending testicular vein suggests that the testis is absent.
- *Diagnostic laparoscopy is very helpful*: The absence of testicular vessels or presence of blind-ended testicular vessels suggests that the testis is absent on that side (agenesis).

II. ECTOPIC TESTIS

An ectopic testis, like the undescended testis, should be placed in the scrotum as soon as the condition is suspected.

In these cases orchidopexy is usually a simple procedure because of the sufficient length of the cord.

Hormonal treatment is of no value.

III. RETRACTILE TESTIS

Retractile testis does not require any treatment. Reassurance to the parents that testicular descent will occur normally (may be delayed upto puberty) is all that is necessary.

Hormonal treatment is of little value.

Comparison between undescended and ectopic testes

Undescended testis	Ectopic testis
1. Arrested descent of the testis which lies in the normal path of its descent.	1. The testis deviates from its normal path of descent.
2. Cause: Already mentioned.	2. Cause: Already mentioned.
3. Undescended testis is poorly developed (cf. retractile testis – the testis is usually of normal size).	3. Usually well developed.
4. Scrotum is not developed and is empty (cf. retractile testis – the scrotum is usually developed but empty).	4. Scrotum is usually developed but empty.
5. Undescended testis lies deep in the inguinal canal (cf. retractile testis).	5. It lies superficial to inguinal canal.
6. On pressure – the testis disappears upward, backward and laterally. It may disappear deep into the abdominal cavity.	6. No effect on pressure.
7. Length of the cord is usually short.	7. Length of the cord is usually adequate.
8. Power of spermatogenesis becomes poor.	8. Power of spermatogenesis – usually normal.
9. Treatment – requires operation and depends on the age and may be satisfactory (cf. retractile testis).	9. Treatment – requires operation.
10. Complications and hazard – so many – see below.	10. The only hazard is liability to injury (rarely the testis may be associated with an oblique, or interstitial inguinal hernia).

Role of hormone therapy

Much controversy exists regarding the efficacy of hormone therapy in truly cryptorchid patients. Most consider it to be efficacious only in retractile testis. It may be used in patients with bilateral palpable cryptorchid (inguinal) testes with or without hypogonadism and obesity.

Drugs used

At present two types of hormones are available: hCG and GnRH (LHRH). hCG is given IM in the dose of 1500 IU/m² on alternate days for 9 doses.

GnRH is given in the dose of 1.2 mg/day as a prenasal spray for 4 weeks.

Drawbacks of hormone therapy

- Injections are painful.
- It may precipitate precocious puberty.
- Mechanical barrier is almost always present in truly cryptorchid testis on which hormone has no effect. Rather it will be a waste of time.
- Inguinal hernia coexists in majority of the cases and if the hernia is not timely treated there is a chance of strangulation.

CYSTS IN RELATION TO EPIDIDYMISS

A cyst of the epididymis may arise due to cystic degeneration of the vestigial remnants in the epididymis. These may include:

- Remnants of the mesonephric or Wolffian duct system:
 - The appendix of the epididymis.
 - The paradidymis.
- Remnants of the para-mesonephric or Mullerian duct system
 - The appendix of the testis (Fig. 11.11A).

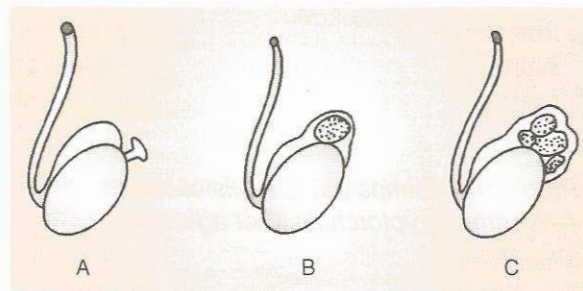


Fig. 11.11. Cysts in relation to epididymis. A, cyst of the hydatid of Morgagni; B, spermatocele; C, cyst of the epididymis.

CYST OF THE EPIDIDYMIS

It is a *multilocular degeneration cyst* in relation to head of the epididymis.

Diagnostic criteria

1. Though congenital in origin, cysts of the epididymis are usually *found during middle life*.
2. Patient complains of swelling in the scrotum. The swelling enlarges very slowly over many years. It may be bilateral.
3. The swelling is situated at the head of the epididymis and hence *above and behind the testis*. The testis can be distinctly felt apart from the swelling (Fig. 11.11C).
4. On careful palpations – *multilocular cystic swellings*; the swelling feels somewhat lobulated or like a branch of tiny grapes (because it consists of an aggregation of small cysts (Fig. 11.11C).
5. *Fluctuation* – positive.
6. *Transillumination* – *brilliantly positive* but is finely tessellated (due to the presence of numerous septae) giving Chinese lantern or marble floor appearance.
7. *On aspiration* – *crystal clear fluid* without any sperm will be obtained.

Treatment

1. If the swelling is small – it does not require any treatment.
2. If the swelling is large enough to cause persistent pain or discomfort—
 - Excision of the cyst completely via a scrotal incision.
 - Disadvantage: Excision will almost certainly cause infertility from blockage and the patient should be informed of it before undertaking the operation.
 - Aspiration of the cysts may be performed but it is useless as the cysts are multilocular and repeated aspirations may introduce infection.

SPERMATOCELE

It is a *unilocular retention cyst* derived from some sperm conducting mechanism of the epididymis, either from the vasa efferentia of the testis or from the epididymis.

Diagnostic criteria

1. A soft cystic swelling situated in the head of the epididymis and hence above and behind the testis (Fig. 11.11B). The testis can be distinctly felt apart from the swelling.
2. Fluctuation – positive.
3. Transillumination: Poorly positive (cf. cyst of the epididymis).
4. *On aspiration* – *white, opalescent (barley water-like) fluid* containing dead spermatozoa is obtained (cf. cyst of the epididymis).

Treatment

1. If of a smaller size – it does not require any treatment.
2. If of a larger size causing discomfort—
 - Aspiration – may be useful as it is a unilocular cyst.
 - Complete excision of the cyst is performed via a scrotal incision.

Differential diagnosis

The *cyst of the epididymis* and *spermatocele*: Both the types of cyst are to be differentiated from a vaginal hydrocele by the following points:

- Both these cysts are situated above and behind the testis. So, they can be distinctly felt apart from the testis. In hydrocele, the testis cannot be felt separately.
- They are softer in feel but hydrocele is a tense cystic swelling.
- Transillumination test:
 - Cyst of the epididymis – positive with Chinese lantern appearance.
 - Spermatocele – poorly positive.
 - Hydrocele – positive but uniform appearance.
- Aspiration test – The aspired fluid:
 - Cyst of the epididymis – crystal clear.
 - Spermatocele – white opalescent (barley-water like).
 - Hydrocele – amber-coloured (urine-like).

PROCESSUS VAGINALIS

Its source and fate (Fig. 11.12)

Processus vaginalis is a peritoneal diverticulum dragged down by the testis during its descent from the abdomen to the scrotum.

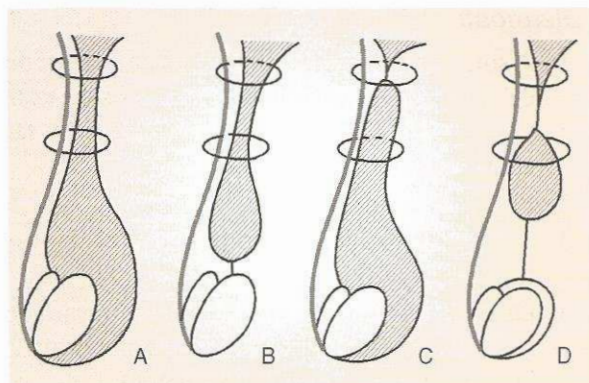


Fig. 11.12. Abnormalities arising in relation to the processus vaginalis. A, congenital hydrocele; B, funicular hydrocele; C, infantile hydrocele; D, encysted hydrocele of the cord.

After the testicular descent is completed, the processus vaginalis becomes occluded at two points soon after birth – first at the deep inguinal ring, and secondly, just above the testis leaving behind a part of the sac in relation to the testis which is known as the Tunica Vaginalis of the testis. The tunica vaginalis surrounding the testis has two layers – the visceral layer and the parietal layer.

The portion of the processus vaginalis between the two occlusions is known as funicular process, which normally becomes obliterated forming a fibrous cord, the rudiment of processus vaginalis.

In some children, this process of occlusion and/or obliteration may not take place and the persistence of the processus vaginalis is the principal factor in the development of congenital hernia and hydrocele.

The difference between a congenital hernia and congenital hydrocele in a child is the size and the contents of the processus vaginalis. A hernia contains an intra-abdominal structure whereas a hydrocele contains only peritoneal fluid.

HYDROCELE

A hydrocele is an abnormal collection of fluid within some part of the processus vaginalis, commonly in the tunica vaginalis of the testis.

Clinically, two types are encountered according to the presence or absence of a communication with the peritoneal cavity.

1. **Communicating hydrocele:** Generally present in neonates and children, and results from the incomplete obliteration of the processus vaginalis. This patent processus vaginalis contributes to the collection of peritoneal fluid in the tunica vaginalis.
2. **Non-communicating hydrocele:** Typically appears in adult life.

Causes of hydrocele

- Congenital
- Acquired
 - Primary or idiopathic
 - Secondary

Primary or idiopathic

The cause is unknown. *There is no associated disease of the testis or epididymis.* It is thought that an imbalance of the secretory and absorptive capacities of the layers of the tunica vaginalis is responsible for the collection of fluid within the sac.

Primary hydrocele usually develops slowly and becomes large and tense.

Secondary

It occurs *due to some pathological condition of the testis or epididymis*, e.g.

- Trauma
- Inflammation – epididymo-orchitis
- Lymphatic obstruction – filariasis
- Tumour – malignancy

Most secondary hydroceles appear rapidly and remain small and lax. Exception is hydrocele due to filariasis which can be large and tense.

Types of primary hydrocele

Five anatomical varieties are encountered:

1. Congenital hydrocele
2. Funicular hydrocele
3. Infantile hydrocele
4. Encysted hydrocele of the cord – rare.
5. Hydrocele of the canal of Nuck.
6. Vaginal hydrocele – commonest

CONGENITAL HYDROCELE

Here the processus vaginalis remains patent throughout communicating with the abdominal cavity and fluid accumulates in it; but the communication at the



Fig. 11.13. Congenital hydrocele – brilliantly transilluminable.

deep ring is too narrow to admit the bowel or omentum; it only admits the escape of fluid (cf. Hernia).

Diagnostic features

1. Congenital swelling present since childhood.
2. It appears as an inguinoscrotal or scrotal swelling which becomes more prominent on standing or walking.
3. The hydrocele volume fluctuates, with the scrotum being full by evening after a day's activity, but relatively empty in the morning after the fluid has drained back into the abdominal cavity through the patent processus during night rest.
4. Testis cannot be felt separately from the swelling.
5. Cystic swelling – fluctuation positive.
6. Cough impulse – present.
7. Reducibility – reducible, partly on lying down and on gentle pressure and this confirms the presence of a patent processus vaginalis; *no gurgling on reduction*.
8. Transillumination – positive.

FUNICULAR HYDROCELE

This is also congenital but largely incomplete, i.e., the processus vaginalis is shut off from the tunica vaginalis just above it.

The features are the same as those of congenital hydrocele except that the testis can be felt separately.

INFANTILE HYDROCELE (Fig. 11.14)

It is a misnomer as it does not indicate a hydrocele in the infant. It is a hydrocele of the sac of the tunica

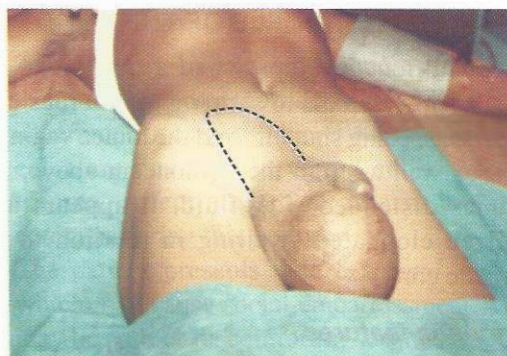


Fig. 11.14. Right-sided infantile hydrocele (also called bilocular hydrocele). Scrotal swelling extending above to the inguinal region upto the mid-inguinal region (note the ink marking over the inguinal region). On examination – cystic swelling, cross fluctuation positive and transillumination positive. Not reducible and cough impulse negative (cf. complete inguinal hernia) (Courtesy: Dr. R. L. Modak, MS)

vaginalis of the testis the upper limit of which extends up to but not beyond the deep inguinal ring. The anatomical funicular process remains as a patent tract instead of being fibrous cord.

Diagnostic features

1. Inguinoscrotal swelling – *Bilocular swelling*, one in the scrotum and one in the inguinal region between superficial and deep inguinal ring communicating with each other.
2. Testis cannot be felt separately.
3. Cystic swelling – *cross fluctuation positive*.
4. Cough impulse – absent.
5. *Not reducible* (cf. Inguinal hernia).
6. Transillumination – positive.

Treatment of hydroceles in infants and childhood

Spontaneous closure and resolution of the patent processus vaginalis usually occur by 12 to 18 months of age. *So, if the hydrocele persists beyond 2 years of age surgical treatment is necessary.*

Operation is performed through a small inguinal crease incision, as the primary abnormality is at the level of the deep inguinal ring. The operation consists of high ligation of the patent processus vaginalis at the deep ring followed by excision of the part of the distal sac. The remaining part of the opened sac is left intact.

ENCYSTED HYDROCELE OF THE CORD

This condition develops when a portion of the funicular process fails to shrink into a fibrous cord and persists, being shut off from the tunica vaginalis below as well as from the peritoneum above, and becomes distended with fluid. It appears as a localized elongated swelling in relation to the spermatic cord.

Diagnostic features

1. The swelling may be found in the inguinal, inguinoscrotal or in the scrotal region depending on which part of the funicular process is patent (Figs. 11.16A and 11.16B).
2. Elongated cystic swelling in relation to the spermatic cord.
3. The swelling is limited both above and below and the testis can be felt apart from it (Fig. 11.12B).
4. Cystic swelling – fluctuation positive.
5. Cough impulse – absent.
6. Not reducible.
7. Transillumination – positive.
8. *Traction test* – positive, i.e., when gentle traction is exerted on the testis, the swelling comes down and becomes fixed or less mobile.

Treatment

Excision of the encysted cyst.



Fig. 11.15. Right-sided encysted hydrocele of the cord – inguinoscrotal swelling. Note that the testis was felt separated from the swelling. The upper margin of the swelling extended to the inguinal canal. Cough impulse negative. Transillumination positive. Traction test positive.

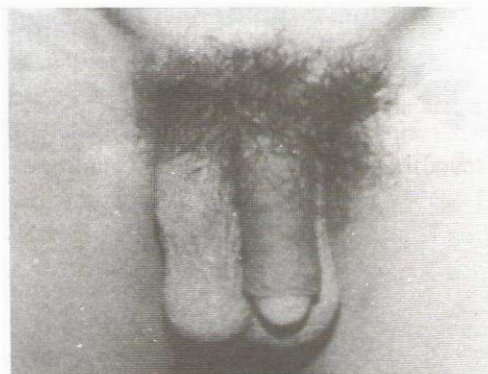


Fig. 11.16A. Right-sided encysted hydrocele of the cord – scrotal swelling. Cough impulse negative. Transillumination positive.



Fig. 11.16B. Same patient. The scrotal swelling was limited both above and below and the testis could be felt apart from it. Traction test positive.

HYDROCELE OF THE CANAL OF NUCK

It occurs in the female rarely, in relation to the round ligament, always in the inguinal canal.

VAGINAL HYDROCELE

Here the fluid collects between the visceral and parietal layers of the tunica vaginalis of the testis. It gives rise to a scrotal swelling.

Composition of hydrocele fluid

Hydrocele fluid is amber or straw-coloured. It contains water, inorganic salts, traces of albumin and fibrinogen. In long-standing cases the fluid may contain cholesterol and tyrosine crystals. The fluid does not clot until it comes in contact with blood.

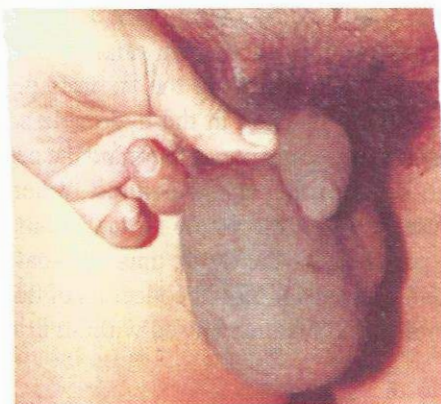


Fig. 11.17. Right-sided vaginal hydrocele. Scrotal swelling. The upper limit of the swelling can be reached – **getting above the swelling** one can feel the cord of the testis above the lump. Fluctuation positive. Transillumination positive.

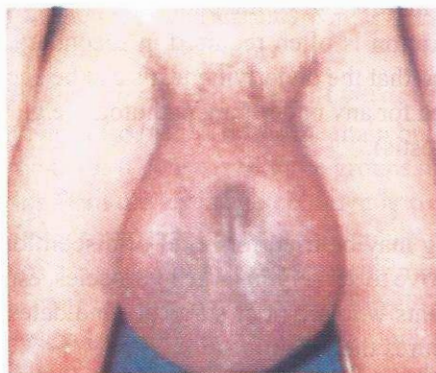


Fig. 11.18. Large bilateral hydrocele with buried penis. Fluctuation positive. Transillumination positive. After eversion of vaginal sacs the penis regained its shape.

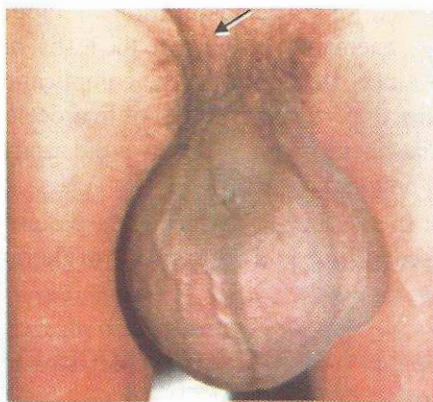


Fig. 11.19. Bilateral hydrocele right > left. Also note the right inguinal swelling – incomplete inguinal hernia. So, always examine inguinal region for hernia while examining for hydroceles and vice versa.

Diagnostic criteria of vaginal hydrocele

1. Age: Primary hydroceles are most common in the middle-aged or the elderly men, but it is **not** uncommon in early childhood. Secondary hydroceles are common in young adults because trauma, infection and neoplasm are most common during this period.
2. The patient presents with a *scrotal swelling* which may cause social embarrassment. There may be pain or discomfort in secondary hydrocele but idiopathic hydrocele can reach a considerable size without causing pain.
3. The swelling may be unilateral or bilateral.
4. The swelling is confined within the scrotum. The upper limit of the swelling can be reached i.e., *one can get above the swelling* (Fig. 11.17) (cf. congenital hydrocele, infantile hydrocele, inguinoscrotal hernia).
5. *One can feel the cord of the testis above the lump.*
6. The testis cannot be felt separately as it is surrounded by a bag of fluid. (cf. encysted hydrocele of the cord, cyst of epididymis).
7. Uniformly cystic – *fluctuation is positive*.
8. The swelling may be tense or lax depending on the amount of the contained fluid.
9. *The swelling is not reducible* (cf. hernia).
10. *Transillumination – positive* as the hydrocele fluid is clear. Transillumination may be negative if the tunica or skin is thick, or the fluid is turbid. (But one must be careful to exclude testicular tumour when transillumination is negative in a scrotal swelling. *Any scrotal swelling when felt hard do transillumination test; if negative, the testicular tumour is suspected.*)

Differential diagnosis

1. Inguinoscrotal hernia.
2. Cysts in relation to epididymis.
3. Tumour of the testis.
4. Chylocele.
5. Haematocele.
6. Filariasis of the scrotum.
7. Encysted hydrocele of the cord.

Complications

1. Rupture
2. Haematocele following aspiration or trauma
3. Infection – may lead to pyocele

4. Atrophy of the testis
5. Hernia of the hydrocele sac through the dartos muscle.
6. Calcification of the sac.
7. Cosmetically unsightly when very big.

Treatment

Indication

1. When the scrotal mass is cosmetically unsightly.
2. Uncomfortable for the patient.
3. If any complication arises.
4. Appears to be a bar to entry into the armed or police service.

Methods

Various methods are available including surgery, aspiration, aspiration followed by injections of a sclerosing agent, e.g., tetracycline within the sac, tapping.

Surgery

Surgery offers the best chance of cure and recurrence is unlikely. It can be performed also under local anaesthesia. Types of operations are:

1. **Jaboulay's operation:** Mostly practised now-a-days for small and medium sized hydroceles.

Principle:

- The vaginal sac is incised anteriorly and the fluid is drained out.
 - The sac is then everted behind the testis and epididymis so that the secreting surface of the tunica vaginalis will be lying outside. The everted margin of the sac is stitched with a running suture.
 - Meticulous haemostasis is achieved.
 - The scrotal wound is closed with or without drain.
2. **Excision of the sac** – may be necessary in a very large hydrocele, haematocele, chronically inflamed hydrocele (e.g., chylocele), as the sac wall becomes thickened in these cases.

After draining the fluid, most of the vaginal sac is excised leaving behind a margin of 2 cm by the side of the testis. Bleeding from the cut margin is always considerable and should be controlled either by continuous mattress suture or with diathermy. The scrotal wound is closed preferably with a drain. Excision of the distal part of the scrotal skin may also be required to reduce the size of the scrotum.

3. **Lord's procedure:** A more or less bloodless operation often practised in small hydroceles. After draining the fluid the testis is delivered through the incision in the hydrocele sac. The edges of the sac are plicated around the periphery with several interrupted sutures. When these sutures are tied, the whole tunica is bunched at the periphery of the testis, thus eliminating the potential space for further collection of fluid. The scrotal wound is then closed without drain.

Aspiration

Aspiration with or without infusion of a sclerosing agent such as tetracycline, is often tried as an alternative to surgery, especially in patients who are poor surgical candidates. But this method is associated with a high rate of recurrence with or without pain in almost half of the patients.

Aspiration is often required in secondary hydroceles, so that the underlying testis can be accurately palpated for any underlying pathology (e.g., tumour of the testis).

Tapping

Tapping may be tried for relief of discomfort as an alternative to surgery in large hydroceles, especially in patients who are poor surgical candidates. But it never cures the condition.

Before tapping with trocar and cannula one must perform the transillumination test which will indicate the site of the testis and scrotal vessels which must be avoided.

Complications after aspiration or tapping are infection, haematocele, injury to the testis, high rate of recurrence. Because of these complications and the ability to perform operation of hydroceles using local anaesthetic, many prefer to avoid aspiration or tapping now-a-days.

Note: In presence of chylocele, the patient should be advised post-operatively a course of Hetrazan 100 mg t.d.s. for 3 weeks.

FILARIAL HYDROCELE (CHYLOCELE)

- Usually occurs following repeated attack of filarial epididymitis.
- Commonly occurs in the coastal regions – southern and eastern states of India.

- Hydrocele is usually large in size and the sac is thickened.
- Usually present as tense cystic swelling – fluctuation is positive.
- *But transillumination is negative* because of thick opalescent fluid content.
- On aspiration or drainage – thick yellowish opalescent fluid is drained (cf. pyocele).
- Fluid content is rich in cholesterol and fat.
- Treatment: Inversion of sac and a course of antifilarial drugs.

VARICOCELE

It is a condition of dilatation, elongation and tortuosity of the veins of the Pampiniform venous plexus. In many cases, it has been found that the testicular veins are normal and the varicosities are present in the cremasteric veins which anastomose freely with the testicular veins.

Surgical anatomy of pampiniform plexus

The pampiniform plexus forms the main bulk of the spermatic cord. It consists of three groups of veins:

1. Veins from the testis and epididymis.
2. Veins accompanying the vas deferens.
3. Veins associated with the cremaster muscle.

Veins of testis and epididymis form an anastomosing plexus surrounding the vas. From the upper part of the plexus 15 to 20 veins arise and as they ascend the number is reduced to about 12. On reaching the superficial inguinal ring, these unite to form 4 veins and traverse the inguinal canal. On reaching the deep inguinal ring these further join to form 2 veins. The veins thereafter enter the abdomen and unite at various levels to form a single vein called testicular vein. The right testicular vein drains in the inferior vena cava at an acute angle. The left testicular vein drains in the left renal vein at right angle. Near the termination the testicular vein is provided with valve (Fig. 11.20).

The cremasteric veins and the vein accompanying the vas anastomose with the testicular veins. So, they provide an alternative pathway (collateral) of venous return from the testis and epididymis. The cremasteric veins drain into the inferior epigastric vein.

Surgical physiology

The pampiniform plexus around the testicular artery is believed to constitute a countercurrent system

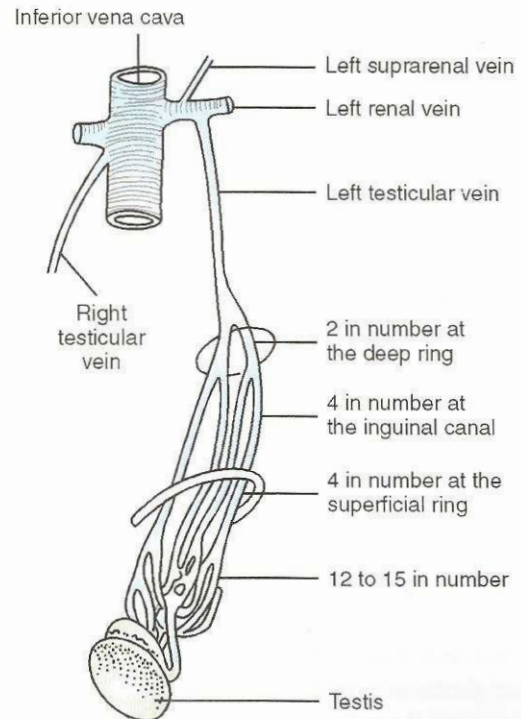


Fig. 11.20. The pampiniform venous plexus and mode of drainage of testicular veins.

which serves as a heat exchanger mechanism to reduce the scrotal temperature to keep the testicle cool and thus, this has some useful role to play in spermatogenesis.

Types of varicocele

1. *Primary or idiopathic:* Where no definite cause has been found. One hypothesis is that it occurs principally in young unmarried men and due to chronic venous congestion consequent upon unrelieved sexual stimulation. Most varicoceles (95%) are idiopathic.
2. *Secondary:* Where there is an obstruction of the testicular vein by some intra-abdominal or retroperitoneal pathology e.g., a tumour in relation to the left kidney.

Aetiology

No definite aetiology has yet been found as the contributing factor in case of primary varicocele.

1. *Age:* Mostly seen in teenaged individuals or in early adult life. Tall, thin, visceroptotic men are frequently affected.
2. *Incidence:* About 10 percent of males between 15 and 25 years have a varicocele.

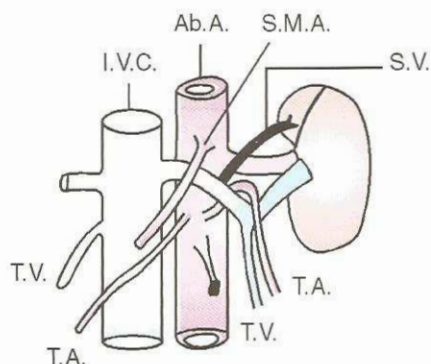


Fig. 11.21. Compression of the left renal vein by superior mesenteric artery (S.M.A.); arching of the left renal vein by left testicular artery (T.A.); close association between the left suprarenal vein (S.V.) and left testicular vein (T.V.). I.V.C., inferior vena cava; Ab. A., abdominal aorta.

3. *More common (95 percent) in the left side for the following reasons:*

- The left testicular vein joins the left renal vein almost at a right angle. The right testicular vein joins the inferior vena cava at an acute angle (Fig. 11.20).
- The left testicular vein is longer than the right because the left testis lies slightly at a lower level and the left testicular vein ends at a higher level. So, the left vein has to bear a long column of blood, and thus the pressure is greater.
- Loaded pelvic colon is likely to compress the left testicular vein as it ascends behind the pelvic colon.
- Sometimes the left renal vein may be sandwiched between the abdominal aorta and the trunk of superior mesenteric artery. Eventually, it hinders the venous return of the left renal vein and therefore of the left testicular vein, which may culminate into varicosities of the left pampiniform plexus (Fig. 11.21).
- Left testicular artery may arch over the left renal vein and thus may cause compression over it (found in 16 percent cases according to Nathan) (Fig. 11.21).
- The close association between the entry of the left suprarenal vein and the left testicular vein in the left renal vein may allow the circulating adrenaline to constrict the testicular vein (Figs. 11.20 and 11.21).

4. There may be incompetency or absence of valves near the termination of testicular veins.
5. Rarely the tumour tissue of renal carcinoma grows along the left renal vein, and thus may cause obstruction to the left testicular vein. So, renal carcinoma is to be suspected when a recent varicocele appears in a middle-aged individual (secondary varicocele).

Power of spermatogenesis in varicocele

Power of spermatogenesis may be seriously depressed (e.g., oligospermia) because the affected testis is subjected to a higher temperature following the stagnation of blood in the varicose veins. It is said that unless the testis is kept at a temperature of 2.5°C lower than the rectal temperature normal spermatogenesis will be hampered. *Varicocele is one of the causes of male subfertility or infertility.* Both the sperm count and motility are reduced. Operative correction will improve both but mainly the sperm motility.

Surgery can result in improvement in fertility up to 50 percent.

Diagnostic criteria

1. Mostly seen in the teenaged individuals or in early adult life. Gynaecologist many a time refers a male patient with subfertility or infertility to exclude varicocele.
2. More frequent and more troublesome in hot climates.
3. It may give rise to either scrotal swelling (elongated scrotum) or inguinoscrotal swelling. The left side alone is involved in 90 percent of cases, with bilaterality in 5 to 20 percent.
4. The patient complains of dragging pain in the affected side which may extend into the groin. (The elongated scrotum no longer supports the testis whose full weight is now borne by the cord giving rise to dragging pain).

Examination:

5. *On palpation with the patient standing:* A mass of dilated, tortuous veins lying posterior to, and above the testis is felt which gives the impression of 'a bag of worms' – chief characteristic. The degree of dilatation can be increased by the valsalva maneuver.

6. *On coughing:* The swelling gives an impulse like a fluid thrill (cf. hernia gives an expansile impulse). Fluid thrill may be absent in secondary varicocele.
7. When the patient lies down and the scrotum is elevated, the swelling reduces in size and disappears as the veins will be emptied by gravity. *During this examination, the size of the testis should be compared with that of the opposite testis.* Testicular atrophy from impaired circulation may be present. The testis on the affected side is usually somewhat smaller and softer than the opposite testis in long standing cases.
8. *On standing:* The swelling reappears, the varicocele fills up from the bottom of the scrotum (cf. Hernia).
9. Fluctuation and transillumination – negative.
10. *Bow sign:* The varicocele mass is held between the fingers and the thumb and the patient is asked to bow down – the tension within the veins becomes appreciably less (because bowing will cut off the continuity of the blood inside the varicocele mass with that of the testicular vein).
11. *Always the abdomen should be examined in case of varicocele particularly in middle aged individuals* – to note a lump e.g., renal carcinoma, loaded pelvic colon.

How to differentiate a primary varicocele from a secondary varicocele?

In primary idiopathic varicocele, on elevation of the scrotum in a patient lying down, the varicocele disappears as the veins will be emptied by gravity. *But in secondary varicocele*, the varicocele will not disappear because either the testicular vein is compressed by a renal lump or the left renal vein is filled up with tumour embolus in case of renal carcinoma, so that it obstructs the drainage of testicular vein. When suspected IVP, ultrasound, CT scan, selective arteriogram will confirm the renal carcinoma.

Investigations

1. Ultrasound of the scrotum to confirm scrotal varicosities.
2. Semen analysis of the male patient with subfertility or infertility – to look for oligospermia, oligoasthenia.

Treatment

A. Conservative

Reassurance and suspensory bandage usually suffice and high Y-front underpants may relieve ache and discomfort.

B. Operative

Indication:

1. Pain and heaviness are constant sources of discomfort and anxiety to the patient.
2. When gross varicosity is present leading to a big size – not relieved even by conservative treatment.
3. When the testis hangs at an abnormally low level.
4. Subfertility (a suboptimal semen analysis e.g., oligospermia, oligoasthenia) or infertility.
5. When varicocele appears to be a bar to entry into the armed or police forces since the condition tends to worsen in hot climates on prolonged standing; this may be a readymade excuse for malingering.

Aim of operation: To reduce to a minimum the number of veins in the pampiniform plexus.

Operative procedures: Different methods are available which account for their own merits.

- Inguinal approach – classical.
- Scrotal approach.
- Abdominal extraperitoneal approach.

1. Inguinal approach

- *Through inguinal approach*, the canal is opened and the spermatic cord is delivered.
- The coverings are incised. The vas deferens together with its artery and one or two veins are carefully separated from the main mass of dilated vessels.
- Then the clamps are applied both above and below the isolated mass of dilated veins with a gap of about 2 inches and the intervening segment is removed.
- The clamps are then approximated and the individual masses are tied off together with a single ligature of strong silk.

Advantage of this procedure is two-fold:

- (a) occlusion of most of the dilated veins, and
- (b) shortening of the cord so that the testis is suspended at a higher level.

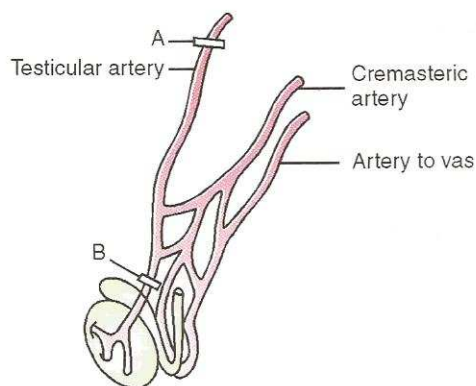


Fig. 11.22. The anastomoses of the testicular artery with cremasteric artery and artery to the vas; ligature of testicular artery at A – still the blood supply to testis is maintained. But ligature at B imperils the blood supply to the testis.

Disadvantage: Chance of damage to the testicular artery and/or cremasteric artery.

2. Scrotal approach

- Through scrotal incision the varicose veins are exposed right down to the level of the testis.
- Tortuous and varicose veins are divided and ligated individually keeping behind sufficient veins to maintain venous return.

Disadvantage of this method: Separation of the veins becomes more difficult near the testis where a free anastomosis exists; bleeding is therefore more troublesome, and a scrotal haematoma is more liable to complicate the convalescence. There is every chance of damage to the testicular artery also.

3. Palomo operation

- A small oblique incision is made 3 cm above the level of the deep inguinal ring.
- Splitting of the parietal muscles is done in 'gridiron' manner.
- Extraperitoneal fat and peritoneum are exposed and they are swept medially, upwards and forward.
- The testicular vessels are exposed as they lie on the posterior abdominal wall, lateral to the external iliac artery which serves as a useful guide.
- The vein is identified, separated and divided between the ligatures.

Advantages of Palomo's method:

- (a) Simple operation, it can be performed under local anaesthesia.

- (b) Less chance of endangering the blood supply to the testis, as the operation is carried out well above the deep inguinal ring.
- (c) Even if the testicular artery is divided and ligated, the testis will get adequate blood supply via the anastomosis between the cremasteric artery and the artery to the vas which cannot be injured at this higher level (Fig. 11.22) (cremasteric artery – a branch of the inferior epigastric artery; artery to vas – a branch of the superior vesical artery).
- (d) Moreover, the cremasteric vein, which anastomoses with the testicular vein and thus provides an alternative pathway (collateral) of venous return from the testis, is not destroyed by this method. So, the testis can be drained by this route to the external iliac vein.

Disadvantage: It aggravates the varicocele in about 10 percent cases.

Laparoscopic ligation

The principle of operation is the same as Palamo's operation. Laparoscopically, the testicular vessels are exposed. The testicular vein is identified and separated. The testicular vein is endoclipped with endoscopic ligacclip applicator. The vein is then divided between the clips with endoscissors.

FOURNIER'S GANGRENE

(Syn.: Idiopathic gangrene of the scrotum)

- It is a variant of synergistic gangrene involving the scrotum due to vascular insufficiency of infective origin (cf. Maleney's superficial gangrene of the abdominal wall).
- It may be idiopathic.
- It may follow minor injuries or procedures in the perineum such as bruise, scratch, dilatation of stricture urethra and drainage of perineal abscess.
- It is common in patients of low socioeconomic groups living in an unhygienic condition.
- In recent times, the disease has been observed in chronic alcoholics who develop pressure sores of the scrotum and perineum after sitting in the same position for a long time in a drunken stupor.
- Diabetic patients are also prone to this condition.



Fig. 11.23. Fournier's gangrene involving both scrotums. (Courtesy: Dr. Sandip Chakravorty, MS, DGO)

Causative organisms

Multiple organisms like:

- (i) *Haemolytic streptococci* (sometimes micro-aerophilic) – gram-positive, associated with
- (ii) *Staphylococci* – gram-positive
- (iii) *E. coli* – gram-negative
- (iv) *Cl. welchii* – anaerobic

At first these organisms set up a fulminating inflammation of the scrotal subcutaneous tissue that results in obliterative arteritis of the arterioles supplying the overlying skin leading to gangrene and necrosis.

The testes covered by tunica and the spermatic cord are usually spared.

Diagnostic features

1. Common in young apparently healthy young men between 25 and 50 years of age.
2. Initially the patient presents with sudden pain of the scrotum with prostration, pallor, and pyrexia. The scrotum becomes inflamed – red, swollen and tender.
3. Within one or two days, extensive superficial gangrene develops resulting in necrosis and sloughing of the scrotal skin. The sloughing may be partial or total extending to the base of the penis.
4. The cellulitis may spread along the fascial planes (so well known as in superficial extravasation of urine) to the perineum and anterior abdominal wall.

The testes and spermatic cord are usually spared.

Treatment

1. **Antibiotic therapy:** Pending the bacteriological report, following antibiotics should be started immediately:
 - (i) Penicillin or Ampicillin – for gram-positive organisms
 - (ii) Chloramphenicol or Gentamycin – for gram-negative organisms
 - (iii) Metronidazole – for anaerobes
 - (iv) Cephalosporin may be added if required particularly after antibiogram.
2. **Surgical debridement of the scrotal skin and subcutaneous tissue:** As soon as possible to provide early drainage and thus to prevent spread of gangrene.
3. If the testicles are exposed, they can be implanted in the thigh.
4. Once the infection and necrosis subside, the wound is covered with split skin graft.

Differential diagnosis of scrotal swellings

While examining a scrotal swelling one should decide first *whether the lump is in the skin of the scrotum* (e.g., sebaceous cyst, filariasis) or not.

Once a scrotal skin swelling is ruled out, one should next decide *whether the swelling is a true scrotal swelling or an inguinoscrotal swelling*. To decide this, one should examine the swelling with the patient standing up and *try to get above the swelling* (Fig. 11.17) to feel the cord of the testis above the swelling.

Getting above the swelling is not possible: Inguinoscrotal swelling (e.g., hernia).

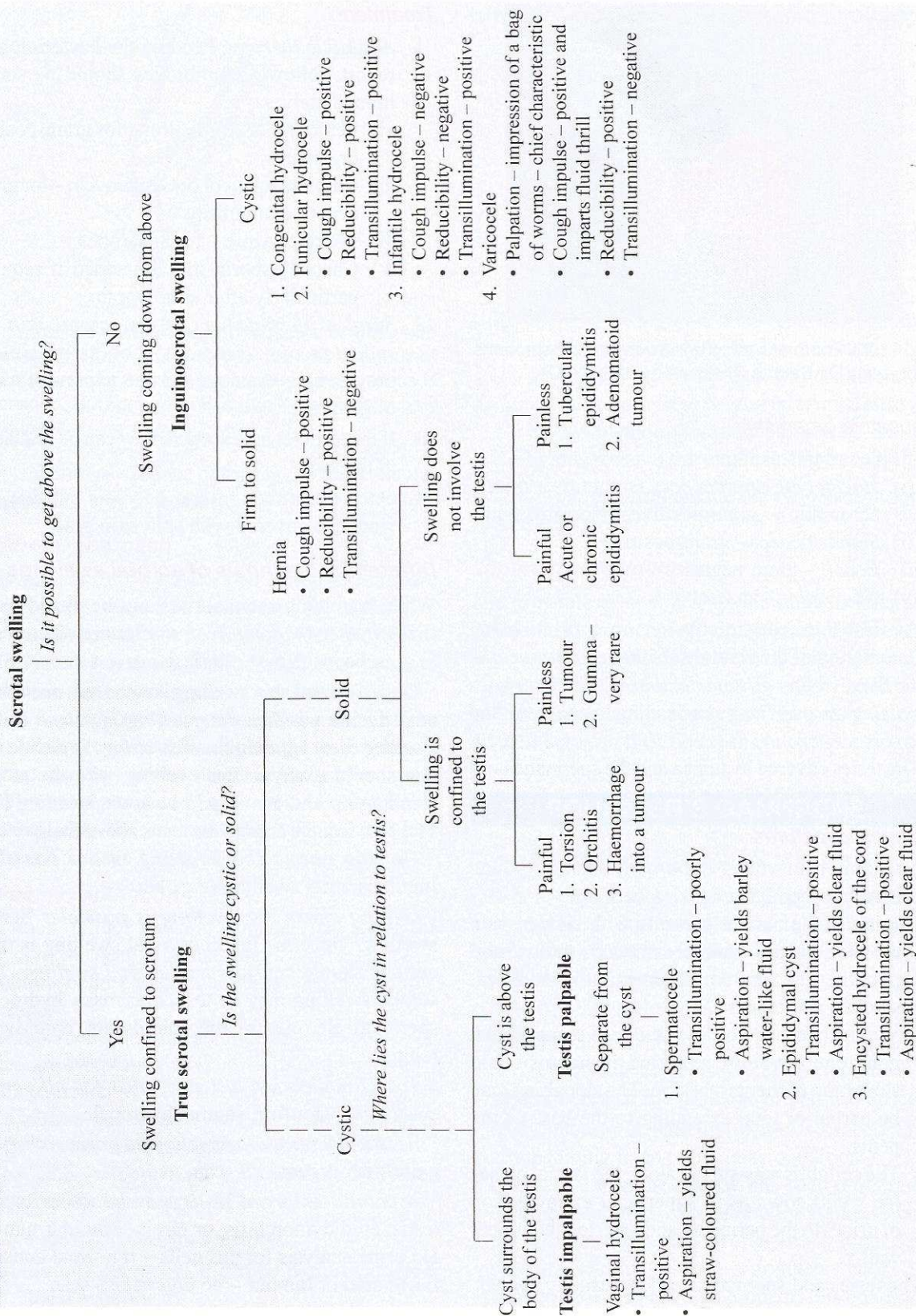
Getting above the swelling is possible: Scrotal swelling. Once the inguinoscrotal swelling is ruled out one should consider *true scrotal swelling*. True scrotal swelling may be testicular mass, hydrocele, spermatocele, cyst of the epididymis, epididymorchitis.

Transillumination test is a must for all scrotal swellings to confirm vaginal hydrocele.

A testicular mass is a tumour until proven otherwise – consider urgent U/S scan.

A painful enlarged testis may be either torsion testis, epididymorchitis, or rarely testicular tumour. Do urine analysis for pus cells – if normal consider for torsion or tumour – do urgent U/S scan.

Algorithm for Examination of Scrotal Swellings



Note: In doubtful cases ultrasound scan solves the problem in most of the cases.

12

CHAPTER



Penis

PHIMOSIS

Phimosis is the narrowing of the end of the prepuce (foreskin) so that it cannot be retracted to expose the glans penis. Because of this, the child may have difficulty in passing urine.

Phimosis should not be confused with redundancy of the prepuce, where the fore skin can be retracted to expose the glans penis.

In neonates and children up to 3 years the prepuce may be normally adherent to glans penis. So it is not possible to expose the glans by retracting the prepuce but the external meatus can always be clearly seen. So, that is not a true phimosis.

Causes

Congenital

Not common.

Acquired

1. Inflammatory: Due to scarring following long-standing infection from poor local hygiene e.g., balanoposthitis, amononical dermatitis. Most cases occur in uncircumscised males.
2. Traumatic: The trauma of forcible retraction causes scarring.
3. Neoplastic: Underlying carcinoma may lead to narrowing of the preputial orifice.



Fig. 12.1. Phimosis. Narrow preputial opening, external meatus not visible. Skin cannot be retracted proximally over the glans.

Diagnostic criteria

1. Babies may be brought by the parents with the complaints of difficulty with micturition, inability to retract the prepuce. There may be history of *ballooning of the prepuce* during micturition – an important criterion of true phimosis.
Young adults may present with the difficulty of retraction of the prepuce causing interference with erection and sexual intercourse.
Patient may present with recurrent balanitis causing pain and purulent discharge.

2. Difficulty in retracting the preputial skin with narrowing of the preputial orifice.
3. There may be associated pin-hole meatus.

Complications

- | | |
|--------------------|---|
| 1. Balanitis | } These may be the causes of phimosis also. |
| 2. Posthitis | |
| 3. Balanoposthitis | |

Because of the inability to clean the glans in a case of phimosis the person is prone to develop recurrent balanitis, posthitis or balanoposthitis due to irritation by the smegma and/or infection. The patient may present with local pain and purulent discharge.

4. Balanitis xerotica obliterans
5. Paraphimosis: This is a condition in which the tight foreskin or prepuce once forcibly retracted over the glans becomes stuck behind the glans penis and cannot be replaced in its normal position. The everted skin fold acts as a constricting band at the corona glandis and results in venous congestion leading to oedema and swelling of the glans, which leads to more difficulty in reducing back the tight prepuce over the glans. The constricting band of preputial skin also becomes oedematous and swollen.

In neglected cases, arterial occlusion and necrosis of the glans may occur.

Emergency surgical relief is mandatory. Following emergency relief, the patient must return back early for elective circumcision.

6. Ballooning of the preputial sac: In extreme cases the preputial sac balloons out with the accumulated urine when the patient micturates and a weak narrow stream of urine flows.
7. Calculus: May form in the preputial sac. Smegma along with urinary salts make such calculus.
8. Obstruction to the flow of urine may cause retention of urine, residual urine, hydroureter, hydronephrosis from back pressure. These are rarely due to phimosis alone; more often there is associated pinhole meatus. *So, check for pinhole meatus in every case of phimosis.*
9. Recurrent urinary tract infection: *Paediatrician many a time refers the children to a surgeon for correction of phimosis.*



Fig. 12.2. Carcinoma of glans giving rise to phimosis. On retraction of the preputial skin proximally exposed the growth.

10. Carcinoma: Phimosis itself is an aetiological factor in the development of carcinoma of the penis. Development of carcinoma has been attributed to the chronic irritation by the smegma which is a product of bacterial action on desquamated cells retained within the preputial sac. *Circumcision carried out soon after birth confers total immunity against carcinoma of the penis.*

On the other hand, carcinoma can give rise to phimosis. So, any old man presenting with phimosis should be examined properly to exclude a hidden carcinoma of the prepuce or the glans penis. Often, a dorsal slit of the prepuce is required to exclude a hidden carcinoma of the glans penis.

Treatment

Circumcision

Excision of the long preputial skin to expose the glans penis and the corona glandis under general or local anaesthesia.

When phimosis is associated with narrow external urethral meatus particularly in congenital variety meatotomy is performed.

When it is associated with considerable inflammation of the prepuce a dorsal slit is performed first, followed by circumcision at a later date when infection subsides.

Complications of circumcision

1. Too little or too much skin excision.

2. Local and/or general infection.
3. Haemorrhage — mostly from the frenal artery.
4. Meatal stenosis.
5. Urethral fistula.

CARCINOMA OF THE PENIS

Carcinoma of the penis commonly occurs in middle-aged and elderly persons. It is uncommon in Europe and USA but relatively frequent in the East, including India.

In 97 percent of cases, it is squamous cell carcinoma and is relatively prevalent among people of low socioeconomic class who have poor health practices and inadequate hygiene.

Sporadic cases of non-squamous malignancy like melanoma, basal cell carcinoma, lymphoreticular tumours, mesenchymal tumours and metastatic tumours have been reported. Rarely, there may be an adenocarcinoma when it arises from the Tyson's gland, the smegma secreting gland on either side of the phrenum.

Aetiology

1. Phimosis: More than 50 percent of carcinoma of penises are associated with phimosis. It has been attributed to the chronic irritation by the smegma collected in the preputial sac. Circumcision carried out soon after birth confers almost total immunity against carcinoma of penis, but circumcision done after early infancy or in adult life does not provide the same degree of protection. The reason is not known. It is almost rare in some communities, e.g. Jewish, where ritual circumcision is performed soon after birth. Moslems, in whom the ritual circumcision is done between the ages of 4 and 9 years, may be affected.
2. Balanoposthitis: Recurrent attacks due to inadequate hygiene may predispose to carcinoma.
3. Papilloma: Condyloma acuminatum or venereal warts of long standing.
4. Buschke-Lowenstein tumour.
5. Leukoplakia of the glans.
6. Paget's disease of the penis.
7. Erythroplasia of Queyrat.
8. Bowen's disease.

9. Balanitis xerotica.
10. Syphilis – doubtful.

Pathology

The lesion usually occurs on the glans and the inner surface of the prepuce near its reflection from the corona. The growth starts either commonly as a wart or an ulcer.

Macroscopic appearance

1. *Ulcerative type* – with rolled out and everted margin.
2. *Proliferative or cauliflower type* – which may be warty or nodular. This type is less malignant than ulcerative type.
3. *Fissure type* – looks like a small fissure situated over the glans.
4. *Plaque type*.

Microscopic appearance

The histologic features of penile carcinoma resemble *squamous cell carcinoma* occurring elsewhere in the body.

Spread

Direct spread: The growth is slowly growing and remains localised for many months. Spread to the body of the penis is prevented for a long time by Buck's fascia surrounding the corpora. Once the neoplasm penetrates the fascia and tunica albuginea of the corporal body, rapid dissemination takes place and vascular dissemination is possible.



Fig. 12.3. Carcinoma penis involving the glans (proliferative type) with induration extending to the shaft. The external urethral meatus was difficult to see, but the patient did not have any difficulty in passing urine. Inguinal nodes were not palpable.

Urethra remains uninvolved even in late cases though scrotal skin may be involved by direct contact.

Lymphatic spread is the earliest: It usually occurs by embolisation through the lymphatics first to the superficial and deep inguinal nodes and then to the iliac lymph nodes. *It is to be noted that penile lymphatic drainage is essentially bilateral.* (Glans penis drains into the *Cloquet's lymph node* which is a deep lymph node in relation to the femoral septum.)

Blood spread: Distant metastases by blood is very rare and commonly involve the lungs, liver and bones.

Diagnostic criteria

1. Age: Common in middle aged and elderly persons. It may also occur in young males below the age of 40 years (40 percent cases).
2. Duration: Usually short and of a few months only. The progress is slow.

Symptoms:

3. (a) The patient usually presents with a lump or an ulcer, representing the papilliferous or ulcerating varieties of penile carcinoma respectively. The lesion as such is a painless condition but may be painful if infected.
- (b) Other symptoms may include mild irritation with penile discharge – serosanguineous, purulent or blood stained.
- (c) The patient may also present with phimosis which may be the cause of carcinoma or may be caused by carcinoma. In these circumstances the primary lesion is hidden under the preputial skin and a dorsal slit is required to expose the lesion.
- (d) Unfortunately many patients delay to report to the doctor early because of embarrassment, guilt, denial, fear or ignorance. They present late with a fungating and foul smelling mass destroying a good portion of the body of the penis.
- (e) The patient may present with enlarged inguinal lymph nodes – which may be secondary to deposits or infections.
- (f) General features of malignancy – Anaemia, anorexia, loss of weight, cachexia are rare.

On examination:

4. Site: The lesion may be anywhere on the glans

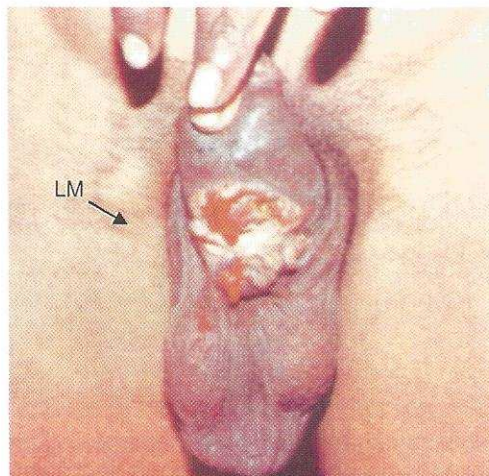


Fig. 12.4. Carcinoma penoscrotal junction ulcerative type – right inguinal nodes were enlarged (stage III).

or the inner surface of the foreskin. The penis may be swollen at the tip due to mass beneath the prepuce. When the tumour is large it may appear through the preputial opening or erode through the preputial skin.

5. *Shape and consistency:* It may appear as an ulcer with a raised, everted edge and necrotic indurated base, or as a sessile papilliferous growth with indurated base. Occasionally, the lesion may be fungating with partial or total loss of the penile shaft. The growth is hard in consistency particularly at the base. The surface of the papilliferous variety is soft and friable and bleeds easily. Any part of the penis which is infiltrated also feels hard. *Entire shaft has to be examined for evidence of tumour infiltration.*
6. *Lymph nodes:* Inguinal lymph nodes are found to be enlarged in about 60 percent cases, but a half of these enlargements is due to secondary infection from the primary growth. Metastatic lymph nodes are hard in feel and may be mobile or fixed. In advanced case, the lymph nodes may appear as fungating growth through the skin of the groin. In very late case, the fungating growth may erode the underlying femoral vein or artery to cause torrential haemorrhage.
7. Urethra is rarely involved in carcinoma penis because it is protected by the Buck's fascia. In a large fungating lesion involving the glans it may be difficult to identify the external urinary

meatus. In such situations, patient will point out the site of exit of urinary stream i.e. the site of external urinary meatus.

Investigations

1. *Biopsy of the lesion and of the lymph nodes.*
Biopsy of the lesion is mandatory prior to therapy even in frankly malignant tumours on clinical examination. Biopsy helps in two ways:
 - (a) Diagnostic value.
 - (b) Therapeutic value – whether radio-resistant or radio-sensitive.

2. Blood for W.R. and Kahn tests.

Importance:

- (a) Syphilitic ulcer and malignant ulcer may co-exist or a malignant ulcer may supervene on a pre-existing syphilitic ulcer.
 - (b) Carcinoma supervening on pre-existing syphilitic ulcer is mostly radio-resistant and hence should be treated by surgery. (*Radio-sensitivity depends on the vascularity of the lesion. Syphilis causes endarteritis obliterans.*)
3. Chest X-ray, lymphangiography, abdominopelvic CT scan, bone scan may be required to note the nodal involvement and dissemination.

Tumour staging

- | | |
|-----------|---|
| Stage I | Tumour confined to the glans or foreskin or both. |
| Stage II | Tumour involves the shaft of the penis. |
| Stage III | Tumour associated with inguinal lymph nodes involvement but operable (Fig. 12.4). |
| Stage IV | Disease is disseminated i.e. the disease is associated with inoperable inguinal lymph nodes or distant metastasis or scrotal involvement. |

Termination of the lesion

1. Haemorrhage – following erosion of the femoral artery or external iliac artery by the fungating metastatic lymph nodes. There may be torrential haemorrhage which often causes death of the patient.
2. Malignant cachexia in late stage, due to:
 - (a) exhaustion from pain, and
 - (b) septic absorption leading to septicaemia.

Treatment

It includes:

1. Treatment of the primary growth.
2. Treatment of the secondary lymph nodes.

TREATMENT OF THE PRIMARY GROWTH

- Surgery
- Radiotherapy
- Chemotherapy
- Cryosurgery
- Laser therapy

The objectives of selecting therapy are to eradicate the primary lesion, prevent local recurrence, and *preserve maximum usable penile length*. In most cases surgery is preferred for management of the primary lesion.

Surgery

Indications

1. Growth larger than 3 cm and invasive.
2. Infiltration of the shaft of the penis.
3. Anaplastic growth.
4. Radiotherapy failed.
5. Carcinoma supervening on leukoplakia or syphilitic ulcer.
6. Elderly males, who do not mind the mutilation as much as the discomfort or pain apprehended from the extensive reaction to radiotherapy.

Types of surgery

1. *Circumcision alone* can be performed for non-invasive tumours involving only the foreskin.
2. *Local excision of the tumour confined to the glans should be avoided* because recurrence rates are high, as much as 40 percent with a worsened prognosis.
3. *Partial amputation*, including 2 cm of benign tissue proximal to the tumour margin is indicated – for growths involving the glans penis or distal shaft, provided the remaining penile length is sufficient to allow to direct his urinary stream from the standing position. This is carried out with the use of long ventral flap, and the urethra is brought out through an opening in it.
4. *Total amputation of penis with perineal urethrostomy.*



Fig. 12.5. Partial amputation of penis for carcinoma penis. 10 months later patient developed metastatic lymph node at left groin – pointed by the tip of the middle finger. Index finger pointed the penile stump.

Indication of total amputation:

- Anaplastic growth.
- Growth involving the proximal shaft or base of the penis.
- Growth in which the extent of disease precludes salvage of sufficient length to allow upright voiding (at least 2 cm of the dependent stump is required).

The procedure includes the removal of the corpora cavernosa from the pubic bones followed by division of the corpus spongiosum leaving at least 1.3 cm of the urethra at the proximal end, which will be taken out through a separate incision behind the scrotum in the form of perineal urethrostomy.

Problem with urethrostomy

- Ammonical dermatitis of scrotum.
- Stricture of perineal meatus – requires repeated dilatation by Hegar's dilator or meatal dilator.

Problem with the scrotum and testes during surgery

There are three views:

- The scrotum and the testes are preserved to avoid psychoneurotic disturbances.
- The scrotum and the testes are removed. The advantage is two-fold.

- The removal avoids disturbance during passing of urine particularly in total amputation.
 - It kills the sexual desire. If the testes are not removed the patient has got the desire but no way of satisfaction after operation because of total amputation or partial amputation leaving a very small stump.
- The scrotum and the testes are preserved but the testes are ablated by radiation to kill sexual desire.

Radiotherapy

Radiotherapy is appropriate only for limited number of cases. Circumcision with or without meatotomy are necessary prior to irradiation.

Indications

- Small lesion less than 3 cm, which is superficial and non-invasive, and located on the glans penis.
- Young patients with small lesions.
- Patients of any age refusing surgery.
- Squamous carcinoma-in-situ (Paget's disease, erythroplasia of Queyrat, Bowen's disease).

Advantage: It preserves the penile structure and function.

Contraindications

- Carcinoma supervening on leukoplakia and syphilis.
- Lesions invasive and larger than 3 cm.

Problems with radiotherapy

- Meagre facilities of radiotherapy, particularly in our country.
- Longer course of treatment.
- Chances of urethral stricture, stenosis, fistula.
- The penis becomes withered, and shrivelled up.
- Chance of post-radiation sterility due to testicular damage.
- There may be erythema and oedema, transient severe pain, ulceration and necrosis of penis as an extensive reaction to radiotherapy that may force cessation of therapy.
- Recurrences are common after radiotherapy.
- Infection accompanying penile carcinoma decreases efficacy of radiotherapy.

Available methods

1. Interstitial: Implantation with flexible radioactive tantalum wires (total dose – 6000 rads in 5 to 7 days).
2. Surface: Radium mould applicators worn either intermittently or continuously (total dose 6000 rads in 7 to 10 days).
3. Megavoltage: Electron beam (total dose 5000 to 5700 rads in 3 to 5 weeks).

Chemotherapy

Topical chemotherapy with 5-fluorouracil cream is only advocated for the treatment of squamous carcinoma-in-situ lesions (Paget's disease, erythroplasia of Queyrat, Bowen's disease).

Systemic chemotherapy has not proved useful even in disseminated disease. Bleomycin, methotrexate, cisplatin, and 5-fluorouracil may be tried in tumours associated with lymph nodes involvement.

Cryosurgery

Recently cryosurgery for carcinoma-in-situ lesions of the penis is tried with encouraging results. Cosmetic results are as good as those achieved with radiotherapy. No recurrence after 5 years has been reported.

Laser surgery

In few sophisticated centres the neodymium: YAG Laser (Photoirradiation) are being tried in carcinoma-in-situ lesion with encouraging results and good cosmetic effect.

TREATMENT OF THE SECONDARY LYMPH NODES

1. **Clinically no palpable nodes:** Bimonthly observation for 3 years. If nodes appear, staged bilateral ilioinguinal block dissection preceded by FNAC of the palpable nodes.
2. **Lymph nodes are palpable but mobile:**
 - (a) Primary lesion is treated.
 - (b) A course of antibiotic is given post-operatively for 3 weeks and the patient be observed:
 - (i) *if the nodes disappear or diminish in size* – suggestive of infective origin. So, no block dissection but bimonthly observation is required for 3 years.
 - (ii) *if the nodes remain unaltered or become more enlarged* – suggestive of malignant involvement. So, block dissection is to be performed preceded by FNAC of the palpable nodes.

Block dissection: It includes removal of the nodes of both the sides; the more affected side is removed first followed by contralateral block dissection at a later date.

Complication – marked lower limb lymphoedema, delayed wound healing.

3. **Lymph nodes are palpable and fixed** – so inoperable. Palliative radiotherapy and/or systemic chemotherapy is given which can cause temporary regression.

The wait and watch policy i.e., removing the inguinal nodes only when they become palpably suspect, has resulted in greater morbidity and mortality. So, some surgeons advocate excision biopsy of sentinel lymph node in clinically non-palpable inguinal nodes – if biopsy is positive an ipsilateral ilio-inguinal lymphadenectomy is performed.

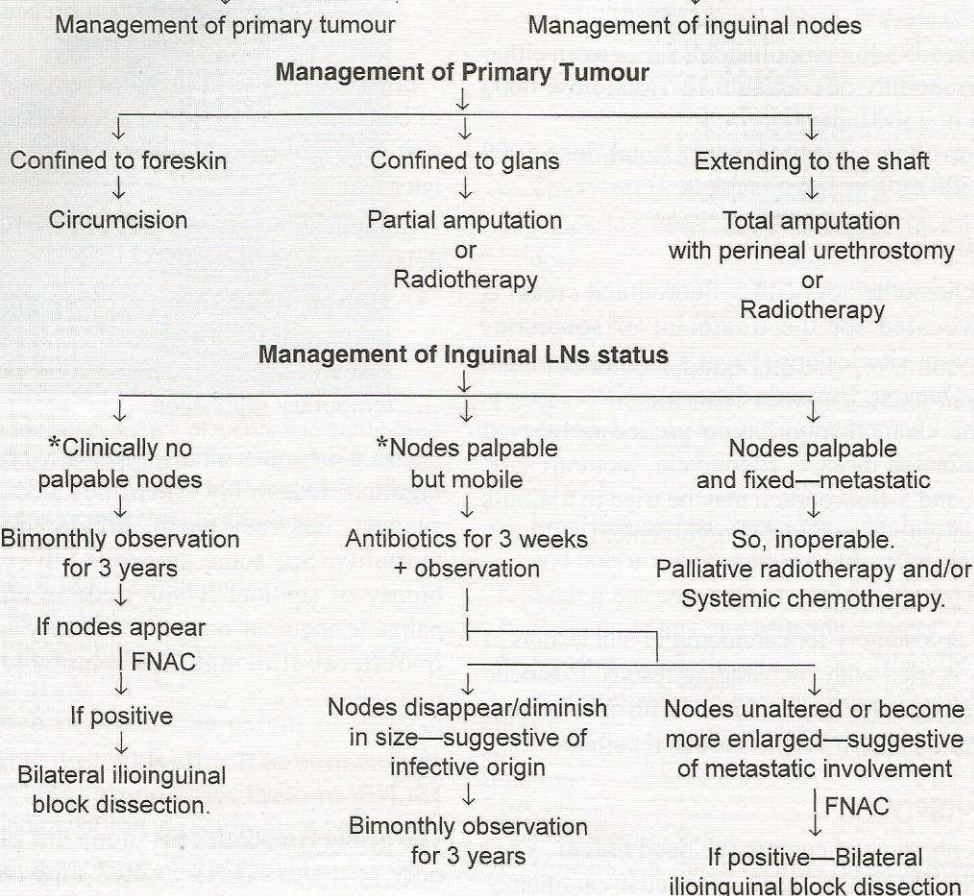
Importance of Sentinel Lymph Node Biopsy (SLNB) in carcinoma penis

The penile lymphatics first drain in a sentinel lymph node or nodes (SLN), located superomedial to the saphenous and femoral vein (saphenofemoral junction) in the area of superficial epigastric vein. *This sentinel node is the first node to get involved in penile malignancy.* In absence of sentinel LN involvement, metastasis to other lymph nodes is most unlikely. So, involvement of SLN (s) will indicate the need for complete ilioinguinal block dissection.

Prognosis

This depends on the site and stage of the growth, and lymph nodes involvement.

- | | |
|-----------|---|
| Stage I | Tumour restricted to the glans penis – 70 percent 5 years survival. |
| Stage II | Tumours involving the shaft of the penis – 60 percent 5 years survival. |
| Stage III | Tumours associated with inguinal lymph nodes involvement – 30 percent 5 years survival. |

MANAGEMENT OF CARCINOMA PENIS

* In either case, without waiting for observation SLNB may be tried and proceed accordingly.

Secondary carcinoma of the penis

- May occur, but rarely.
- Primary source of the lesion is usually the prostate, bladder or rectum.

Way of spread – Three ways:

- by direct spread.
- by retrograde spread.
- by retrograde venous embolism via the dorsal vein of the penis.

PREMALIGNANT LESIONS OF THE PENIS**Condyloma acuminatum (Fig. 12.6)**

1. Rarely occurs before puberty.



Fig. 12.6. Condyloma acuminata – a precancerous lesion.

2. Caused by a papovirus.
3. Transmitted by sexual contact.
4. These are mostly situated near the coronal sulcus.
5. These appear as sharp pointed red, papillary excrescences, sessile or pedunculated and usually moist with an offensive discharge.
6. Can involve the urethra in 5% of cases.
7. They are precancerous lesions.
8. *Treatment:*
 - (a) 20 percent solution of podophyllin in tinct. benzoin application.
 - (b) 0.1% Bleomycin application.
 - (c) Circumcision and fulguration of the lesion.

Buschke-Lowenstein tumour (verrucous carcinoma) (Fig. 12.7)

1. A giant variety of condyloma acuminatum.
2. It appears as large exophytic lesion.
3. It penetrates and destroys adjacent tissue.
4. It is a benign lesion with a pre-malignant potential.
5. Because of its large size it frequently is grossly indistinguishable from squamous cell carcinoma.

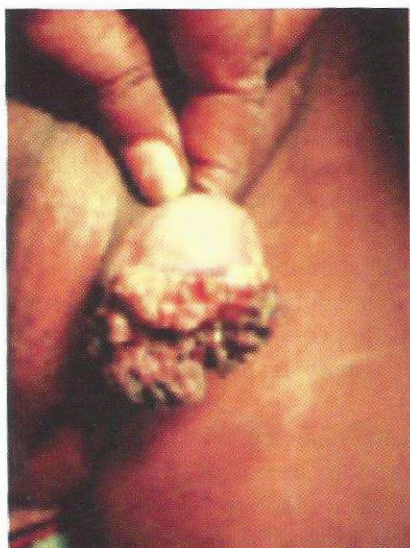


Fig. 12.7. Buschke-Lowenstein tumour. A large exophytic growth on the undersurface of glans nearly obscuring the meatus. Wedge biopsy performed – reported condyloma.

Excisional biopsy or multiple deep biopsies are necessary to exclude true penile carcinoma.

6. *Treatment:*
 - (a) For small lesions – Local excision of the tumour that spares penile anatomic structure, so function is adequate.
 - (b) Large lesions – may require partial amputation of the penis.
 - (c) Bleomycin topically may be helpful.

Leukoplakia of the glans

1. It is a condition of hyperkeratosis of the epithelial cells (cf. tongue, vulva and vagina).
2. It appears as greyish white plaque.
3. The condition is painless.
4. Pre-malignant.
5. *Treatment:* Local excision.

Erythroplasia of Queyrat (Fig. 12.8)

1. It appears as a dark red, velvety, well-margined, flat or slightly raised indurated patch on the glans penis, or coronal sulcus. Occasionally it becomes superficially ulcerative when there may be discharge and pain.

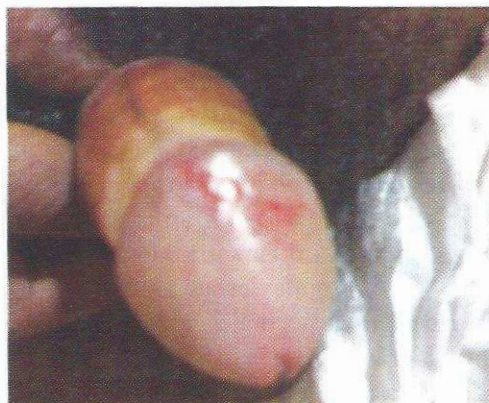


Fig. 12.8. Erythroplasia of Queyrat – a premalignant condition.

2. It is similar to Bowen's disease of penis on histology and represents carcinoma-in-situ changes.
3. Prior to initiating therapy multiple biopsies of adequate depth should be taken.
4. Pre-malignant.

5. Treatment:

- (a) Local excision of the lesion is adequate.
- (b) If a lesion is preputial – circumcision will suffice.
- (c) 5-Fluorouracil cream topically is helpful. Cosmetic results are excellent.
- (d) Nd-YAG laser fulguration is also adequate.

Bowen's disease

1. It is similar to erythroplasia of Queyrat histopathologically and clinically but the lesion is

drier in appearance and is often crusted and ulcerated.

2. Pre-malignant.

Balanitis xerotica

1. It presents as white atrophic lesion with or without oedema involving the glans or prepuce or both.
2. Pre-malignant.
3. *Treatment:* Local excision and/or topical steroid application.

13

CHAPTER



Filariasis and its Surgical Manifestations

Filariasis is tropical disease prevalent in the Southern and Eastern States of India.

It is caused by the infestation with the parasite, Wuchereria bancrofti. It is a nematode transmitted to human beings by a mosquito bite (Culex fatigans).

Wuchereria bancrofti: This parasite passes its life cycle in two hosts – man and mosquito. Man is the definitive host; Mosquito is the intermediate host. The stages are:

1. Adult filariae both male and female are found in the lymphatic system of man.
2. After sporulation, the females die and innumerable embryos or microfilariae are produced.
3. These microfilariae enter the peripheral blood particularly at night (nocturnal periodicity) where they can live for a considerable time but do not undergo any metamorphosis until they pass through the mosquito.
4. When a mosquito bites an individual, it sucks in the microfilariae from the peripheral blood.
5. These microfilariae in the mosquito undergo further development and maturation, but they are still sexually immature.
6. These mature microfilariae again enter a man by the bite of the infested mosquito and ultimately reach his lymphatic system where they settle down. The most favourite site for this settling is the inguinoscrotal region. In the lymphatic system they begin to grow into adult

forms and again the cycle continues.

Pathological changes

The pathological changes are brought about by the adult filariae, living or dead. The microfilariae usually do not produce any pathological changes. The changes may be classified into two parts – lymphangitis and lymphatic obstruction.

Causes of lymphangitis

1. Mechanical factor – irritation of the lymphatic system due to the movements of the adult filariae along the lymphatic system.
2. Allergic factor – *due to helminthic toxins* which are liberated in three ways:
 - (a) Liberation of toxins by the fertilised females during parturition.
 - (b) Liberation of toxins from the dead worms undergoing degeneration.
 - (c) Sometimes in highly reacting individuals, the mild toxins liberated by the microfilariae can produce some allergic reaction.
3. Secondary infection – due to the associated bacterial infection, usually streptococci, often acts as an additional factor, particularly in presence of lymphatic stasis.

So, the toxins liberated give rise to foreign body reaction. Clinically this is manifested by transient attacks of lymphangitis.

Effects of recurrent lymphangitis

1. Proliferation of endothelium of lymphatics and thereby causing occlusion of the lumen – obliterative endolymphangitis.
2. Fibrosis of the lymph vessels in varying degrees. As a result of fibrosis adult filariae are imprisoned in the lymph nodes and in the lymph vessels.

Causes of lymphatic obstruction

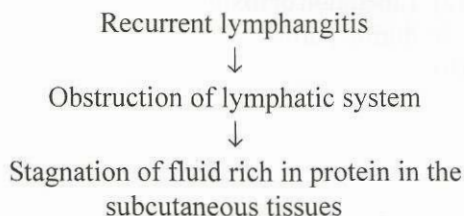
1. Mechanical factor – imprisonment of the adult filariae, both living and dead, in the lymph nodes and lymph vessels. This will produce further foreign body reaction leading to further fibrosis.
2. Sclerosing lymphangitis – leading to fibrosis of the lymph nodes and lymph vessels.

Effects of lymphatic obstruction

I. The lymphatic obstruction will lead to dilatation, elongation and tortuosity of the lymphatic systems giving rise to:

1. Lymph varix or lymphangiectasis – dilatations of the lymph vessels.
2. Lymphadenovarix.
3. Lymph scrotum – dilatation and tortuosity of the cutaneous lymphatics of the scrotum.
4. Hydrocele – due to obstruction of lymphatics there is effusion into the tunica vaginalis.

II. Lymphatic obstruction along with subsequent fibrosis will lead to a condition known as 'Elephantiasis'. *Changes of elephantiasis are brought about in the following way:*



This will cause an increase in the tissue colloidal osmotic pressure which increases filtration of fluid across the capillary membrane and decreases reabsorption with the result *that fluid of a high protein content collects in the subcutaneous tissues* – the affected part becomes swollen, oedematous, and *pits on pressure*. When the attack subsides the swelling is reduced to some extent, but the normality is seldom attained.

With subsequent attacks, the swelling further increases in size and becomes more firm due to excessive growth of connective tissue as a result of stimulation by the protein in fluid. The lymph stasis provides the suitable nourishment for the luxurious growth of the fibroblasts. The subcutaneous tissue is replaced by fibrous tissue and lymph-logged gelatinous tissue known as blubbery tissue (blubber means jelly-fish). *Ultimately, firm, non-yielding and non-pitting swelling is produced.*

The skin becomes enormously thickened and hyperkeratotic with abundant fibrosis under it. The surface of the skin becomes uneven and rugose with warty growth and looks like the skin of an elephant (*hence the name, elephantiasis*). There is also loss of skin appendages e.g., scanty hairs.

Common sites of involvement of elephantiasis:

1. Scrotum
2. Penis
3. Lower limb – usually unilateral
4. Vulva
5. Breast – The glandular tissue of breast remains unaffected
6. Arms.

The scrotum and the lower limb are the common sites for elephantiasis probably because of the following reasons:

- (a) Great laxity of the subcutaneous tissue and the dependent position of the scrotum.
- (b) Dependent position of the lower limb.
- (c) The microfilariae and adult filariae have special predilection for their site to the lymphatics of the inguinoscrotal region.

III. Obstruction and rupture of lymph vessels: Lymphatic obstruction leads to dilatation of the lymph vessels and when the tension is high, they rupture. Either chyle or lymph escapes depending on whether the obstruction is situated above or below the cisterna chyle. The results are:

1. Chyle may come out with urine – Chyluria.
2. Chyle may come out with faeces – Chylous diarrhoea.
3. Chyle may collect in different serous cavities of the body such as:
 - (i) tunica vaginalis – Chylocele
 - (ii) peritoneal cavity – Chylous ascites
 - (iii) pleural cavity – Chylothorax.

ELEPHANTIASIS OF THE SCROTUM

Diagnostic criteria

1. In the early stage, the patient may come with scrotal swelling with pitting oedema, and in advanced case present with solid oedema.
2. The skin is thickened, rough, rugose, fissured and hyperkeratotic. There is loss of hairs. The thickening commences at the most dependent part of the scrotum and extends upwards. In an established case the thickening is maximum at the bottom and minimum at the root of the scrotum.
3. Over the surface of the skin there may be appearance of vesicular eruptions which may ulcerate and discharge clear lymph or milky lymph.
4. Due to excessive thickening with unyielding nature of the scrotal wall it is sometimes impossible to palpate the contents within, viz., testis, epididymis, spermatic cord, etc.
5. At the root of the scrotum and over the inguinal canal – The spermatic cord is thickened on palpation and may be tender.
6. A hydrocele is usually associated, but fluctuation and transillumination are difficult to elicit through the thickened skin.
7. Elephantiasis of the penis may sometimes be associated with elephantiasis of the scrotum. The skin and subcutaneous tissue of the penis become hypertrophied.
Sometimes the penis is enormously thickened and distorted owing to unequal contraction of the hypertrophied fibrous tissue and fascia around the penis. Such a swollen and distorted penis is called '**Ram horn penis**'.
At other times, the penis, being healthy, is buried either completely or partially within the hypertrophied scrotal mass, when its orifice is indicated by the hypertrophied prepuce.
8. Inguinal lymph nodes may be affected, more often in males. The lymph nodes become enlarged and tender.
9. There may be associated involvement of the lower limb or breast.

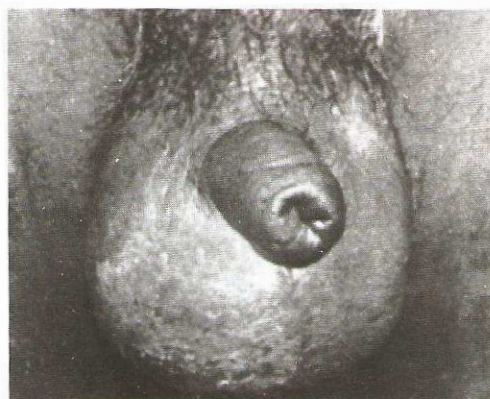


Fig. 13.1. Early stage filarial scrotum and penis.

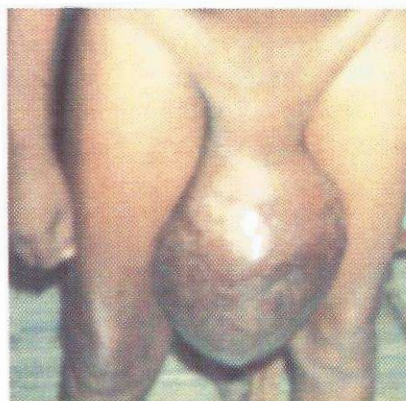


Fig. 13.2. Filarial scrotum with buried penis. (Courtesy: Dr. Yogesh Salphale, D'ortho)



Fig. 13.3. Filarial scrotum with Ram horn penis.



Fig. 13.4. Filariasis involving the labia majora right side. Also note the lymphoedema right leg. This patient had the excision of the filarial swelling of the left labia majora 5 years ago. (Courtesy: Dr. Subrata Biswas, MD)



Fig. 13.5. Filariasis involving the right labia majora.

10. *History of filarial fever is present.* The epididymo-orchitis, funiculitis or acute inguinal lymphadenitis often precedes the development of elephantiasis.

Characteristics of filarial fever

- (a) Periodic in nature i.e., filarial inflammation occurring on full and new moon (not every full and new moon).
- (b) Onset: Sudden with chill and rigor (cf. Malarial fever, fever of urinary tract infection: In both these conditions there will be absence of swelling and tenderness of the spermatic cord).
- (c) Temperature: Often high (103 °F to 104 °F) and may continue for several days (usually 3 to 5 days). The temperature comes down by crisis with profuse sweating.
- (d) Presence of toxic features: Tachycardia, flushed face, dry coated tongue, severe headache and loss of appetite.

- (e) *Simultaneously there may be associated pain, swelling and tenderness of the spermatic cord and scrotum* (these features help in differentiation from malarial fever and urinary tract infection). There is also an associated inflammation of the inguinoscrotal lymph nodes.
- (f) Routine blood examination reveals a transient leucocytosis with an increase of eosinophils.

Investigations

1. (a) Blood for routine examinations: Eosinophilia (5 to 15 percent) is very often noticed in early cases. There may be no eosinophilia in cases of lymphatic obstruction.
- (b) Midnight blood for microfilariae: Thick film in Leishman's stain.

Why midnight blood is drawn:

Microfilariae appear periodically in the peripheral blood in the night. The cause is not known definitely. The probable explanations are:

- (i) It is due to night-biting habit of the vector (*Culex fatigans*).
 - (ii) It is the habit of the person concerned who sleeps at night.
 - (iii) It is the periodic and cyclical emptying of the gravid uterus of matured adult worm.
 - (iv) Sunlight repels the microfilariae (not accepted by all).
2. Immunological test with *D. immitis* antigen and complement fixation test – little value.
 3. Detection of microfilariae in the hydrocele fluid or in the exudate of lymph varix.
 4. Biopsy of the lymph nodes may reveal adult filaria within them.

Treatment

The treatment of choice is surgery. The patient should, however, be prepared preoperatively.

Pre-operative treatment

General:

- (a) A course of specific antifilarial drug – Diethyl carbamazine citrate.
Dose: 6 mg per kg of body weight in three equal doses.
- (i) Hetrazen – 2 tablets thrice daily for 3 weeks.

- (ii) Benocide Forte – 1 tablet thrice daily for 3 weeks.

During the carbamazine therapy *an anti-allergic drug should be given* to combat any allergic reaction caused by the dead microfilariae.

- (b) Antibiotics: If necessary (when routine blood shows polymorph nuclear leucocytosis also) for combating concurrent streptococcal infection.
- (c) Non-specific protein therapy to increase the immunity: Sterodin or Milk injection.

Local:

Treatment of the skin condition e.g., if lymphoria is present: *local dressing with lotio Goulard* or *lotio Acriflavin* (1 in 4,000).

Surgery

Principles of surgery

1. Excision of the hypertrophied and oedematous scrotal and/or penile skin and subcutaneous tissue.
2. Resurfacing of the decorticated penis.
3. Management of the exteriorised testes.

Problems of surgery

1. Increase in size with increase in vascularity: blood transfusion should be arranged to combat blood loss during operation.
2. Injury to urethra – can be avoided by passing Foley's catheter perurethra during operation.
3. Shortage of skin to house the testes – when the testes are placed in the pockets made into the upper thigh.

Essence of operation

1. Dealing with the decorticated penis: The raw shaft of the penis is covered with either the free skin graft taken from the thigh or the preserved inner layer of prepuce or both.
2. Dealing with the exteriorised testes: The problem can be dealt with in two ways—
 - (a) The testes can be covered either by the remnant of scrotal skin or by approximating the skin from the two sides of the perineum.
 - (b) If there is lack of scrotal or perineal skin, pockets in the subcutaneous pouch are made in the medial side of upper part of the thigh superficial to fascia lata, and the testes are then placed in the pockets of the respective sides.
3. The wound is closed with a drain at its lowest angle which should be kept for 48 hours.
4. A catheter (Foley's catheter) should be passed and kept in position for three weeks in order to prevent contamination of the wound with urine.

Why the testes are placed superficial to fascia lata of thigh

In order to avoid pressure imparted by the deep fascia particularly during walking or running which may cause pain and pressure atrophy of the testes.

Why the testes are not placed in the inguinal subcutaneous pouch

Scrotal temperature is 89 °F. The temperature in the subcutaneous pouch in the thigh is also 89 °F as compared with 98 °F in the abdomen or inguinal subcutaneous pouch. So, to avoid atrophy, testes are placed in the thighs.



14

CHAPTER

Raynaud's Syndrome (Syn. Raynaud's Phenomenon)

Raynaud's syndrome is defined as recurrent, episodic attack of vasospasm of the digital blood vessels resulting in **triphasic colour changes** of the digits i.e. pallor → cyanosis → rubor in that particular sequence (remember mnemonics P C R).

This phenomenon affects the fingers and hands most commonly, though it may occur in the feet, ear, nose and lips.

Causes

1. Primary or idiopathic variety: *This variety is known as Raynaud's disease.*

In this condition the digital vessels appear to be healthy but very much sensitive to the cold leading to *vasospasm*. There is no demonstrable structural change in the arterial tree or associated systemic disease. *Raynaud's disease refers to occurrence of Raynaud's phenomenon due to vasospasm without an underlying disease of the digital vessels.*

2. Secondary variety: In this condition Raynaud's syndrome occurs due to obstruction of blood flow in the digital vessels *complicating a known disease or local trauma* e.g.,

- (a) *Collagen diseases* – causing vasculitis
 - Scleroderma – commonest
 - Polyarteritis
 - Rheumatic arthritis
 - Systemic lupus erythematosus (SLE)
 - Dermatomyositis
- (b) *Obliterative arterial diseases*
 - Thrombo Angitis Obliterans (TAO)
 - Atherosclerosis
- (c) *Occupational and industrial exposure to vibrating tools* such as riveting machines and earth impactors which disturb the neurovascular control in the hands and fingers, particularly the pacinian corpuscles.
- (d) *Irritation of nerves and vessels*
 - Thoracic outlet syndrome such as cervical rib, scalene syndrome
 - Cervical disc protrusion
 - Spinal cord disease
 - Old poliomyelitis
- (e) *Peripheral embolism arising from*
 - Damaged subclavian artery crossing a cervical rib
 - Subclavian aneurysm secondary to cervical rib
 - Atherosclerotic stenosis of the subclavian artery
- (f) *Blood abnormalities*
 - Cold agglutinins
 - Cryoglobulins

Both cause vascular damage due to agglutination of red cells e.g., in multiple myeloma.
- (g) *Ingestion of drugs*
 - Ergotamine preparations
 - Methysergide
 - β -blockers, e.g., propranolol

- Amphetamine
 - Cytotoxic drugs
- (h) Poisoning by lead and arsenic
- (i) Atrophic changes in the limb following poliomyelitis, frostbite, etc.

So, basically Raynaud's syndrome may be divided into two distinct pathophysiologic disorders:

- (a) **Vasospastic disorder** in primary variety or Raynaud's disease.
- (b) **Obstructive disorder** in secondary variety.

Sympathectomy is avoided in secondary variety of Raynaud's syndrome as it produces very poor results.

RAYNAUD'S DISEASE

It is a vasospastic disorder of the apparently healthy digital blood vessels in response to abnormal hypersensitivity to cold resulting in alternate vasoconstriction and vasodilatation of the digital arterioles.

The disease was first described by Raynaud in 1862.

Pathophysiology

The exact cause of this arteriolar vasoconstriction is not known. It may be due to:

- (a) Hypersensitivity of the arterioles themselves to cold. The vessels show no evidence of organic disease.
- (b) Abnormality of the sympathetic nervous system α -adrenergic receptors.

The disease is usually precipitated by exposure to cold and/or emotional stress with an abrupt onset.

It passes through three stages and is associated with *cyclical triphasic colour changes* in the digits, and often disturbances of sensation. To remember the colour changes remember the mnemonics W B C (White Blood Cell).

- W White or pale (Stage of blanching)
 B Blue or cyanosis (Stage of dusky anoxia)
 C Crimson or red (Stage of red engorgement)

Three pathological changes are encountered for these triphasic colour changes:

Stage of local syncope: On exposure to cold the digital arterioles go into spasm resulting in diminished blood flow locally which is evident by

pallor or blanching of the fingers (*stage of blanching*). The fingers become cold and waxy pale white. These changes start at the tips of fingers and gradually spread to the base of the fingers. There is also associated tingling and numbness.

Stage of local asphyxia: As the hand is warmed or the general body temperature is raised the spasm relaxes and the capillaries are slowly dilated and filled up with blood. Then as the red cells reach the anoxic tissue they become acutely deoxygenated. The digital vessels gradually become filled up with this deoxygenated blood so that the digits become cyanosed or blue (*stage of dusky anoxia*). The digits are still cold and numb.

Stage of recovery: With further relaxation of the spasm the blood flow is increased and the digits become crimson or red (*stage of red engorgement*), hot and swollen. The redness is also due to a reactive hyperemia set up by the local tissue metabolites which have accumulated during the cold phase. *There is also an acute pain which is tingling and burning in nature.* The pain is due partly to increased tissue tension and partly to irritation caused by local metabolites.



Fig. 14.1. Note the colour changes of the fingers (both the dorsum and palmar surfaces of the fingers) – Raynaud's disease.

Late features

In the early stages of the disease the digits are normal between the attacks but as the disease progresses, obliterative changes may occur in the peripheral vessels – stenosis and/or occlusion followed by *trophic changes in the skin and nails of the fingers*. The patient may eventually get *rest pain and dry gangrene of the finger tips*.

Diagnostic criteria

1. *Sex*: Raynaud's disease is *ten times more common in women than in men*.
2. *Age*: It *usually occurs in young adult* after puberty and before the menopause.
3. In about 50 percent cases there is a family history.
4. It affects the digits of the upper extremity more commonly and more severely than of the lower.
5. Initially one or two fingers are affected but gradually all the fingers are involved to include the distal palm. The thumbs are usually spared.
6. *Commonly bilateral*.
7. *Usually symmetrical*.
8. On presentation, the fingers and hands may look white, and feel cold. Or they may look normal between the attacks. *An attack can be precipitated by asking the patient to dip her hand into ice-cold water*, when the fingers and hands become waxy pale white. If this white hand is then put into warm water, it turns blue or cyanosed and finally a dusky red.
9. In late cases, there may be the presence of trophic changes when the diagnosis can be guessed correlating the patient's description of her typical symptoms with the local findings. *The trophic changes* may be one of the following:
 - (a) blue permanently ischaemic finger tips;
 - (b) thin and pointed finger tips due to atrophy of the pulp;
 - (c) necrosis of small areas of skin followed by small scars;
 - (d) small very painful ischaemic ulcers on the finger tips.
 - (e) recurrent infections around the nails (paronychia) are common. These are painful and slow to heal.
 - (f) the patient may eventually present with gangrene of the finger tips and rest pain.

10. *Pulses*: At the wrist and above are usually palpable. *Presence of peripheral pulses, yet obvious ischaemic changes of the digits is a typical findings in Raynaud's disease*.
11. The return of capillary circulation after blanching produced by pressure on the pulp or nail bed may be very slow.
12. Exclude the causes of secondary Raynaud's phenomenon e.g., cervical spondylosis, cervical rib, scleroderma, history of drug ingestion etc.

Investigations

Diagnosis of Raynaud's disease is made on the typical history and findings.

Special investigations may be performed to rule out any associated systemic disorders leading to secondary Raynaud's syndrome to come to a diagnosis of Raynaud's disease by exclusion.

1. X-ray of the cervical spine AP and lateral views – to exclude cervical rib or spondylosis.
 2. Angiography to establish the diagnosis by:
 - (i) demonstrating the presence of spasm of terminal arterioles on exposure to cold.
 - (ii) Demonstrating subclavian artery stenosis or dilatation, thrombi or emboli.
 3. Doppler ultrasound: To detect the presence of peripheral pulses. It has outdated angiography.
 4. ESR
 5. Rheumatoid factor
 6. Antinuclear antibody
 7. Cold agglutinins
 8. Cryoproteins
- } Rheumatic arthritis
 } Raised in multiple myeloma

Treatment

The treatment of primary Raynaud's disease remains unsatisfactory. There are currently no specific means for overcoming the local fault and the hypersensitivity of the digital arteries to cold.

Conservative

1. Avoid exposure to cold.
2. Avoid any emotional stresses.
3. Avoid smoking or tobacco in any other form.
4. The patient is advised to keep her hands and body warm – to dress warmly, to wear socks and gloves, to keep the house and specially bed warm at night during winter and cold weather. *Warmth of the central body induces peripheral vasodilatation*.

5. Avoid drugs such as ergotamine, methysergide, β -blockers, amphetamine. Avoid smoking.
6. In presence of nail bed infection – oral antibiotics and antifungal drugs like griseoflavin or fluconazole may be advised.
7. Analgesics and neurotonics to give immediate pain relief.
8. Drug therapy to improve vascularity.

(i) *Vasospastic antagonistic drugs*: Drugs that block sympathetic vasoconstriction: reserpine, guanethidine, α -methyldopa, phenoxybenzamine, pentoxifylline and α -blockers like prazosin may be useful.

(ii) *Vasodilators*: Arlidine, complamina, nifedipine (calcium channel blockers). Nifedipine 10 mg BD is commonly used now-a-days.

(iii) I.V. Prostaglandin E and prostacycline are potent vasodilators and inhibit platelet aggregation.

Patients with spasmodic Raynaud's syndrome respond better than obstructive Raynaud's syndrome with drug therapy.

(iv) *Ketanserin* is a selective serotonin receptor which antagonises serotonin-induced vasoconstriction and inhibits platelet aggregation. This drug is useful in treating Raynaud's syndrome induced by scleroderma.

9. Drugs used during acute severe spasmodic attack:

- (i) Intravenous low molecular weight dextran may alleviate the attack.
- (ii) Intra-arterial injection of reserpine (1 mg in 10 ml normal saline) injected slowly in brachial artery may alleviate the attack.

Operative

- Sympathectomy
- Amputation

Cervicodorsal sympathectomy

Procedure

In this operation the followings are removed.

- (i) The portion of sympathetic trunk that includes the lower 1/3rd of the stellate ganglion (containing T_1 ganglion), T_2 and T_3 ganglion which contain most of the postganglionic fibres supplying the upper limb.

(ii) All rami communicantes associated with the T_2 and T_3 ganglion.

(iii) The nerve of Kuntz, gray ramus which arises from the T_2 ganglion and goes to the 1st thoracic nerve.

The stellate ganglion is identified as it lies on the neck of the 1st rib. It is dumbbell-shaped with upper larger inferior cervical ganglion and lower smaller T_1 ganglion fused together with a waist between them. The post-ganglionic fibres supplying the head and neck pass through the inferior cervical ganglion present in the upper larger part of the stellate ganglion.

Removal of the whole of the stellate ganglion will cause Horner's syndrome.

Routes

Cervicodorsal sympathectomy can be done through the supraclavicular route or transthoracic route via an axillary incision at the third intercostal space.

Thoracic route has the advantage of:

- (i) Cosmetically better because of hidden scar at axilla, particularly in female.
- (ii) Removal of sympathetic chain upto T_5 ganglion is possible.

Laparoscopic sympathectomy is being tried at some centers recently.

Result

- The results of cervicodorsal sympathectomy is unpredictable and short lasting.
- The patient may derive some benefit, if it is done in the early stages. In the later stages when the vessels are partly or completely occluded by fibrosis or thrombosis sympathectomy is useless.
- The operation will not provide a permanent cure, as there is no evidence that sympathetic abnormality is only responsible for the disease.
- The maximum result that can be achieved from such operation is some permanent increase in warmth of the digits due to reduction in tone of the cutaneous arterioles.
- There is also some increase in thermal insulation around the digital arteries which can provide a protection against local cooling to critical levels.
- *But in over 75% patients the symptoms return with increased vasomotor tone within five years of operation.*

- The results of lumbar sympathectomy for Raynaud's syndrome in the lower limb are much better and late relapses rarely occur.

Limitations

Sympathectomy should be avoided in secondary causes of Raynaud's syndrome as it produces very poor results.

Complications of cervicodorsal sympathectomy

- Perforation of pleura leading to pneumothorax.
- Haemorrhage leading to retropleural haematoma.
- Lymph fistula following damage to the thoracic duct during left cervical sympathectomy.
- Horner's syndrome – when total stellate ganglion is removed.

Features of Horner's syndrome:

- (i) Ptosis
- (ii) Myosis
- (iii) Enophthalmos
- (iv) Anhydrosis of the ipsilateral face

Indications of sympathectomy

1. Raynaud's disease
2. Cervical rib syndrome when associated with vasomotor disturbances.
3. TAO – Thromboangitis obliterans
4. Herperhydrosis
5. Causalgia
6. Acrocyanosis.

Amputation

Local amputation may be required rarely in cases when there is necrosis of tissues and gangrene of the digits. Since the circulation of proximal vessels of the hand is usually satisfactory, major amputations are seldom required. Any amputation should be supplemented by prior sympathectomy.

ACROCYANOSIS

- This is a chronic persistent vasospastic condition of the cutaneous arteries and arterioles producing cyanosis or blue discoloration of the hands and feet including digits.
- Common in young females.
- There is *persistent* and *painless* cyanosis and coldness of the hands and feet (cf. Raynaud's disease – which is *episodic* and *painful*).

- The cyanosis may also be noticed at the tip of the nose and ears. Often legs are involved.
- The involved parts are also cold with excessive perspiration.
- The peripheral pulses are normal.
- The cyanosis may aggravate on exposure to cold.
- Though the condition is painless (cf. Raynaud's disease), it is susceptible to chilblain. But the digits are not subjected to gangrene.

Treatment

- (a) Conservative: Reassurance and avoidance of undue exposure to cold which can cause chilblains.
- (b) Oral vasodilator drugs: α -adrenergic blockers or calcium channel blockers.
- (c) Sympathectomy in the presence of severe cyanosis or the chilblain is particularly troublesome. The effect may be transient.

THORACIC OUTLET SYNDROME

The thoracic outlet syndrome has a host of synonyms depending on the variation of normal anatomy or the presence of abnormal structures at the thoracic outlet. The synonyms are:

- Cervical rib syndrome
- Scalenus syndrome
- Superior thoracic aperture syndrome
- First thoracic rib syndrome
- Costoclavicular syndrome
- Pectoralis minor syndrome
- Hyperabduction syndrome

All these syndromes give rise to symptoms arising due to neural or vascular compression or neurovascular compression.

Surgical Anatomy

Thoracic outlet is a limited space bounded by the bony structures all around:

- *Anteriorly* – manubrium sterni
- *Posteriorly* – vertebra
- *Laterally* – first rib.

At the root of neck, the *brachial plexus* formed by the five spinal nerves (C_5 - C_8 and T_1) and the *subclavian artery* pass through the scalene triangle (Fig. 14.2). Distal to the scalene triangle at the outer

border of the first rib this neurovascular bundle passes through the costoclavicular space between the first rib and the clavicle. Then the neurovascular bundle enters the axilla behind the insertion of pectoralis minor muscle onto the coracoid process.

So, there are three potential areas of constrictions:

1. In the neck – scalene triangle
2. Costoclavicular space – between the first rib and the clavicle
3. In the axilla – behind the insertion of pectoralis minor.

The syndrome results due to neural or vascular compression or both neurovascular compressions at one of these potential areas of constrictions. However, neural compression is the common presentation and comprises 90% of the symptoms and signs.

CERVICAL RIB SYNDROME

(Syn.: Scalene syndrome, Superior thoracic aperture syndrome)

This is a condition characterised by pain, paraesthesia and muscular wasting of the upper extremity due to some abnormality at the scalene triangle, commonly due to a cervical rib protruding in the scalene triangle.

Boundaries of scalene triangle (Fig. 14.2)

- *Anteriorly* – Scalenus anterior muscle
- *Posteriorly* – Scalenus medius muscle
- *At the base* – First rib.

The trunks of five spinal nerves (C_5 – C_8 , T_1) forming the brachial plexus and the subclavian artery pass through this triangle behind the scalenus anterior muscle. The subclavian vein passes over the first rib

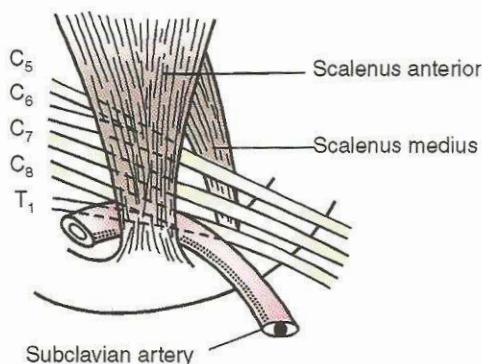


Fig. 14.2. Scalene triangle and brachial plexus.

in front of the scalenus anterior muscle. At the scalene triangle, the neurovascular bundle may be stretched or angulated by one of the following factors:

1. Abnormalities of the rib (Fig. 14.3)

Usually an extra rib springs from the 7th cervical vertebra – the *cervical rib* (cervical rib syndrome). The rib may be complete or incomplete.

- (a) A complete cervical rib – Articulates with the manubrium sterni or first rib (Fig. 14.3A). The scaleni muscles may be attached to such a rib.
- (b) An incomplete cervical rib—
 - (i) The free end of the rib expands and forms a large bony mass (Fig. 14.3B).
 - (ii) The rib may end in a tapering point which is connected by a fibrous band to the scalene tubercle of the first rib (Fig. 14.3C).

Cervical rib is seen only in 1% of the populations, and is bilateral in 80% of cases. Only 10% of the cervical ribs give rise to symptoms.

2. Sometimes, there is no extra rib; only a fibrous band is present extending from the seventh transverse process to either the first rib or scalenus medius muscle (Fig. 14.3D).

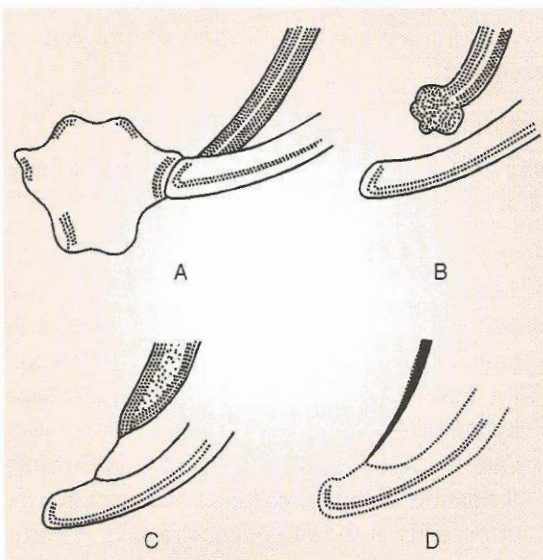


Fig. 14.3. Different types of cervical rib. A, Complete rib articulates with the first rib; B, Incomplete rib ends in a large mass; C, Incomplete rib ends in a tapering point which is connected by a fibrous band to the first rib; D, No rib but only a fibrous band.

3. Occasionally, long transverse process of the 7th cervical vertebra may act as a cervical rib.
4. In absence of a cervical rib, if the first thoracic rib is too wide, the similar symptoms may arise (*first thoracic rib syndrome*).

In all the above mentioned conditions, the neurovascular bundle climbs over the abnormal structure and becomes stretched or angulated.

Abnormality of the muscle

- (a) There may be a close proximation of the insertions of the scalenus anterior and scalenus medius.
- (b) There may be an additional scalene muscle.
- (c) Sclaneus anterior muscle may be wide and thick.

Angulation of the first thoracic nerve

When the brachial plexus is post-fixed, the first thoracic nerve becomes stretched or acutely angulated over the first rib.

All the above mentioned abnormalities are congenital and may not produce any symptoms throughout life; or if produced, the symptoms are usually rare before the age of 30 years. The probable explanation is that with the decline of youth there is gradual sagging of the shoulder girdle, perhaps in association with some atrophy of the regional musculature.

Pathology

There may be angulation or compression of the artery and/or of the first thoracic nerve.

Artery (Fig. 14.4)

- (a) The subclavian artery is rarely compressed, rather it is angulated. Owing to the elevation and angulation, there is constriction or narrowing of the lumen of the artery over the rib – followed by post-stenotic dilatation.
- (b) Due to traumatic arteritis, there is thrombus formation within the post-stenotic dilatation and/or possibly at the site of constriction. A portion of this thrombus may become dislodged and carried distally giving rise to an embolus or emboli of the digital arteries and ultimately gangrene, particularly, of the tips of the fingers.

Rarely, the thrombus may extend proximally and

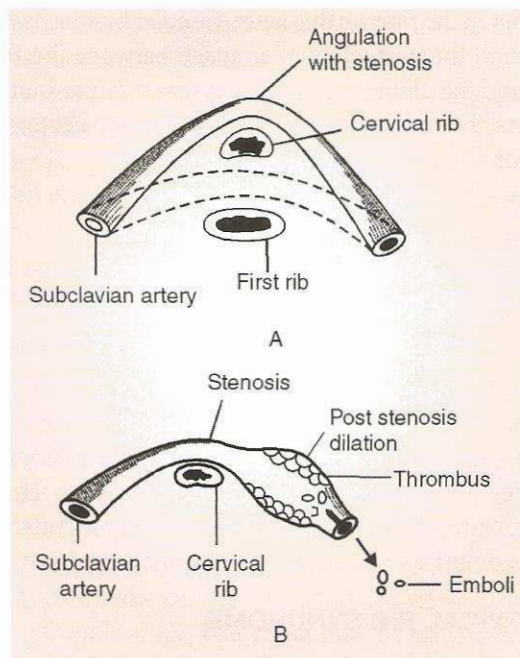


Fig. 14.4A & B. The changes of the sub-clavian artery in relation to cervical rib.

involve the vertebral artery and thus cerebrovascular embolic episodes may occur.

Nerves

- (a) Following stretching or compression of the first thoracic nerve (or the lowest trunk) there will be friction neuritis causing at the beginning sensory disturbances and later also the motor disturbances along the distribution of C₈ and T₁ fibres.
- (b) Irritation of the peri-arterial sympathetic fibres, or paralysis of the sympathetic fibres contained in the lower trunk, or compression of the axillary artery between the heads of a stretched median nerve – may be responsible for the vasomotor disturbances.

Diagnostic criteria

Common in women around the age of 30 years. A thin woman with a long narrow neck is more susceptible.

Symptoms

- Usually, there are no general or neck symptoms. Rarely, dull aching pain in the neck may be caused by expanded bony end of the cervical rib.

- The patient may come with the symptoms depending on the structures compressed. Neural symptoms are the commonest. Arterial symptoms are observed frequently. Venous symptoms are rare.

Neural compression symptoms: Initially result in pain, paraesthesia (pins & needles) principally in the little and ring fingers innervated by the ulnar nerve (C_8-T_1). Later, it may be felt in the ulnar side of the hand and forearm. Often the pain becomes worse after carrying a heavy weight e.g. shopping basket or ironing.

Arterial compression symptoms: Results in pain, numbness, coldness and weakness. Pain is the prevailing prominent symptom, usually an aching pain. The pain is located in the hand and forearm and may radiate to upper arm. Characteristically the pain is brought on by the excessive use of the arm; the onset of pain is accelerated when the arm is in a raised position at the time of exercise. The pain is relieved by rest. *This pain is ischaemic muscle pain similar to intermittent claudication pain of the leg.* In late cases pain is also felt during rest.

Vasomotor symptoms: May be noted as coldness of the fingers and hand, periodic colour changes from the pale to blue (cyanosis), increased sweating, *trophic changes* with ulceration and rarely gangrene of the tips of fingers. *These symptoms are akin to typical Raynaud's phenomenon and are uncommon. Moreover, the symptoms are usually unilateral (cf. Raynaud's disease which is bilateral).* The symptoms appear towards the end of the day or at night and are improved by raising the limb.

Venous compression symptoms: Rare and present as heaviness due to oedema, pain, cyanosis in the hand.

Signs

- Neurovascular signs: Can be grouped under three headings:
 - Signs of neural compression
 - Signs of arterial compression
 - Signs of venous compression
- Signs in the neck

Neurovascular signs

Signs of neural compression

1. Signs of neural compression are less frequent though neural symptoms are common. Usually

features of sensory or motor deficits in the distribution of ulnar nerve may be present:

- (a) Wasting of the small muscles of the hand supplied by the ulnar nerve (C_8-T_1) i.e., interosseous and hypothenar muscles are affected. Hollows between the metacarpals on the dorsum of the hand indicate atrophy of the interosseous muscles.
- (b) Loss of tone and muscle power: Movements of the fingers are clumsy and slow.
- (c) Rarely, there may be clawing of fingers.

2. *The following are the tests to confirm ulnar nerve paralysis:*

- (a) Paralysis of interosseous muscles—
 - (i) Patient is asked to spread out (abduct) and close (adduct) the fingers. Patient will be unable to do it (dorsal interossei are abductors (DAB) and palmar interossei are adductors (PAD)).
 - (ii) **Card test:** Patient is asked to hold a thin paper or card in between the fingers: the patient is unable to hold the card tightly due to paralysis of palmar interossei.
- (b) Paralysis of the first palmar interossei and adductor pollicis:

Book test (Froment's sign): Patient is asked to hold a book firmly between the thumbs and other fingers of both hands tightly. In the normal side the thumb remains straight whereas in ulnar nerve paralysis the terminal phalanx of the thumb of the affected side will be immediately flexed (because in presence of paralysis of palmar interossei and adductor pollicis, the flexor pollicis longus (supplied by median nerve) takes upper hand and flexes the terminal phalanx of the thumb in its attempt to hold the book firmly.

- (c) Paralysis of adductor pollicis muscle – Due to paralysis of this muscle, the patient is unable to hold a piece of paper between the thumb and palm.

Signs of arterial compression

1. Coldness of the fingers, pallor, cyanosis, increased sweating.

2. Rarely, evidence of trophic changes of the fingers is noted – atrophy of the skin, pulp wasting, brittle nails with or without ulceration.

3. More rarely, finger tip necrosis or gangrene is noted. These are due to migration of emboli arising from the thrombus at post-stenotic dilatation of subclavian artery.

4. Feeble radial pulse.

5. *Adson's test*: This is a test to determine the presence of subclavian artery compression.

Feel the radial pulse of the patient sitting on a stool. Now the patient is asked to take a deep breath and hold it, and then the patient is instructed to turn his chin up and to the affected side as much as possible. If the pulse disappears or becomes feeble—indicates the presence of compression of subclavian artery.

Signs of venous compression

Very rare:

1. Distended superficial vein mainly evident on the dorsum of the hand and forearm.
2. Oedema of the hand.

Signs in the neck

1. A palpable lump in the supraclavicular region:
 - (a) The lump may be mushroom like, finely bosselated, hard and fixed bony mass (due to expanded bulbous free end of the cervical rib).
 - (b) The lump may be pulsatile (the subclavian artery being elevated by the abnormal rib).
 - (c) On auscultation over the supraclavicular region – a bruit may be heard.
2. Lowering of the shoulders on the affected side with evidence of muscular wasting.

Investigations

1. X-ray of the neck and chest: May show presence of
 - (i) a cervical rib or long transverse process of the 7th cervical vertebra;
 - (ii) anomalies of the first rib – bifid or wide;
 - (iii) clavicular deformities e.g. excessive callus formation following fracture healing;
 - (iv) narrowing of intervertebral foramina by disc protrusion or exostosis.
2. MRI: May be useful:
 - (i) to identify the brachial plexus and angulation or compression of the lower trunk of brachial plexus;
 - (ii) to identify the cervical disc protrusion and

narrowing of the intervertebral foramina by disc or tumour.

3. Electromyogram and nerve conduction study: To show prolongation of conduction in presence of nerve compression.
4. Arteriogram: Not performed now-a-days because of noninvasive colour Doppler ultrasound.
5. Colour Doppler ultrasound study to know:
 - (i) the exact location of the arterial compression, aneurysm and presence of any thrombus;
 - (ii) the site of venous obstruction.

Differential diagnosis

Many disorders resemble cervical rib syndrome particularly when they are present with neurological features. These are:

1. Cervical spondylosis
2. Carpal tunnel syndrome
3. Ulnar nerve lesions
4. Lesions of the spinal cord
5. Pancoast syndrome
6. Brachial neuritis
7. Raynaud's disease.

Treatment

Before any treatment is undertaken other causes of pain and paraesthesia in the upper limb should be eliminated (see differential diagnosis).

Conservative

1. Postural exercises of the muscles of the shoulder girdle to strengthen the muscles and to lessen the tendency of the shoulder to drop.
2. Analgesics and neurotonics to give immediate relief of pain.
3. Avoid heavy weight-lifting like shopping basket or iron etc.
4. Local heat therapy by ultrasound or short-wave diathermy.

Operative

Indication:

1. Failure of conservative treatment
2. Presence of severe pain and paraesthesia
3. Wasting of the muscles of the hand
4. Evidence of vasomotor disturbances

Essentials of operation:

1. Division of scalenus anterior muscle and/or
2. Complete excision of the abnormal rib or the fibrous band.

These procedures may not cause relief of symptoms. So some surgeons advocate to divide all the three sides of the scalene triangle i.e. division of scalenus anterior and medius muscles, excision of cervical rib or any fibrous band, a portion of the first rib. When a cervical rib is excised, it should be removed along with its periosteum; otherwise there will be a chance of regeneration.

3. If vasomotor symptoms are prominent – the above mentioned operation should better be supplemented with sympathetic denervation of the upper limb i.e., cervical sympathectomy where the 2nd and 3rd thoracic ganglions as well as the lower 1/3rd of stellate ganglion are removed.

Costoclavicular syndrome

This syndrome is due to the compression of the subclavian artery between the first rib and the clavicle.

This leads to the features of arterial compression i.e. pain or paraesthesia, coldness of the fingers, periodic changes in skin colour from pale to blue (cyanosis), trophic changes with ulceration, necrosis and gangrene of the tips of the fingers.

Costoclavicular test

Patient's radial pulse is felt and the patient is asked to brace his shoulders backward and downward as in seen in exaggerated military position—if the pulse becomes feeble or impalpable the test is positive.

Treatment

Excision of the middle 1/3rd of clavicle or the portion of the 1st rib below the subclavian vessels.

Pectoralis minor syndrome

(Syn. Hyperabduction syndrome)

This syndrome is due to the compression of the subclavian or axillary artery beneath the pectoralis minor muscle onto its insertion to the coracoid process. This leads to features of arterial compression.

Hyperabduction test

Patient's radial pulse is felt and then the arm is passively hyper-abducted with or without external rotation – if the pulse becomes feeble or impalpable, the test is positive.

Treatment

Through axillary approach, the pectoralis minor muscle is divided from its insertion onto the coracoid process.

Pancoast syndrome

This syndrome comprises the following components:

- *Pancoast tumour – a peripheral lung carcinoma arising at the apex of the lung.*
- Invasion of the lower roots of brachial plexus (C₈T₁) resulting in symptoms and signs in the distribution of ulnar nerve.
- Invasion of the cervical sympathetic chain leading to Horner's syndrome.
- Erosion of the first rib – as evident on X-ray.
- Common in elderly men who are chronic smokers.

15

CHAPTER



Aneurysm

An aneurysm is defined as a localised dilated sac filled with blood in direct communication with the lumen of an artery.

Causes

Every aneurysm is caused by the weakening of the wall of the artery. The weakness may be congenital or acquired:

1. Congenital

- (a) Berry aneurysm which occurs in the cerebral blood vessels particularly in the circle of Willis is due to congenital deficiency of the elastic lamina at the sites of branching. These aneurysms are usually asymptomatic but may rupture giving rise to subarachnoid or intra-cerebral haemorrhage.
- (b) Congenital arteriovenous fistula where there is a direct shunt between an artery and a vein without the interposition of capillaries. Common in the limbs.
- (c) Cirroid (Racemose) aneurysm – Tortuous complex of dilated arteries and veins. Common in the scalp.
- (d) Aneurysm of the aorta may occur proximally to a coarctation.
- (e) Marfan's syndrome and Ehlers-Danlos syndrome are rare inherited connective tissue disorders and may cause dissecting aneurysm of the aorta.

(Marfan's syndrome is characterised by excessive height, thin body habitus, aortic aneurysm, valvular regurgitation, ocular lesions and skeletal abnormalities.)

2. Acquired

- (a) Traumatic: The wall of the artery may be damaged by a trauma either single or repeated and the weakened wall may subsequently distend to form aneurysm.
Example:
 - (i) Subclavian aneurysm may be related to a cervical rib.
 - (ii) Penetrating wound to the artery may cause an aneurysm – common in limb arteries.
 - (iii) Arteriovenous fistula may develop after trauma.
 - (iv) Radiation injury may cause aneurysm.
- (b) Degenerative: Atherosclerosis, arteriosclerosis, arteriomegaly, cystic medial necrosis. Atheroma is the commonest cause of aneurysm, commonly abdominal aorta. Cystic medial necrosis is the common cause of aortic dissection.
- (c) Infective:
 - (i) Specific: Syphilis now-a-days relatively rare; thoracic aorta most commonly involved.
 - (ii) Non-specific: Mycotic aneurysm due to

bacterial infection e.g., subacute bacterial endocarditis.

- (d) Arteritis – particularly polyarteritis.
- (e) Microaneurysm – associated with hypertension and affects smaller arteries and arterioles especially in the brain.

Sites

Aneurysm may involve both large and small arteries:

- (a) Aorta – Arch of aorta, descending thoracic, abdominal aorta.
- (b) Carotid, subclavian, axillary, femoral and popliteal arteries.
- (c) Smaller vessels like cerebral, mesenteric, renal and splenic arteries.

Types

1. True aneurysm
2. False aneurysm
3. Arteriovenous fistula.

TRUE ANEURYSM

A true aneurysm is due to actual dilatation of an artery, either symmetrical or eccentric. It may be fusiform, saccular or dissecting in nature. The sac wall is lined by the layers of the arterial wall.

Fusiform aneurysm

There is a uniform expansion of the entire circumference of the arterial wall in all directions along the long axis of an artery (Figs. 15.1A & 15.2).

Saccular aneurysm

There is an expansion of a part of the circumference of the arterial wall (Figs. 15.1B & 15.2).

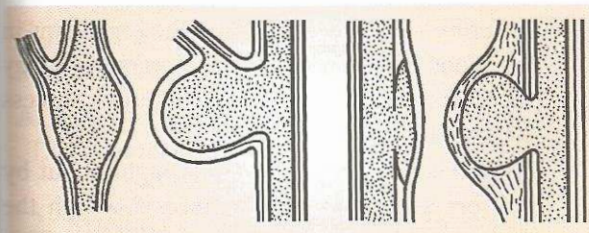


Fig. 15.1. Different types of aneurysm. A, fusiform; B, saccular; C, dissecting; D, false.

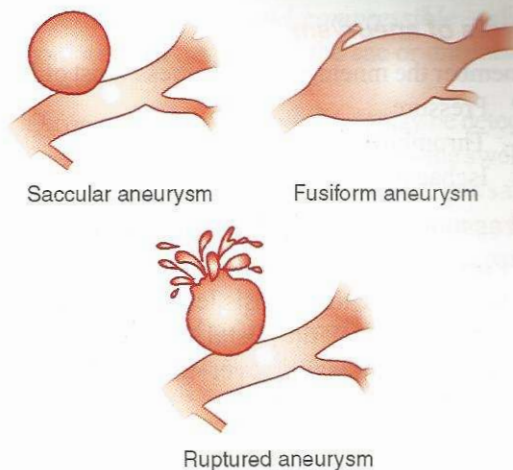


Fig. 15.2. Different types of aneurysm and common complication.

Dissecting aneurysm

A dissecting aneurysm is one in which the blood dissects its way along a tunnel between the medial coat and the adventitia permitting longitudinal propagation of blood-filled space within the aortic wall. It is not true aneurysm, because the vessel is not dilated, but, rather, a haematoma of the arterial wall (Fig. 15.1C).

The leakage of blood may occur at the site of an atheromatous ulcer of the intima, or at the site where the intima has been weakened either by mucoid degeneration or by obliteration of the vasa vasorum.

It usually occurs in elderly hypertensive patients and affects commonly the ascending aorta or near the origin of subclavian artery. With the onset of leakage the patient experiences a sharp pain and a tearing sensation in the chest and there is a severe shock. If the lesion is untreated, death occurs within 24 hours in 50 percent cases. Rarely, the false channel tunnels back into the proper lumen of the aorta and spontaneously relieves the condition.

FALSE ANEURYSM

A false aneurysm is a sac formed by condensed periarterial fibrous tissue and it communicates with the lumen of the artery through an opening in its wall (Fig. 15.1D). *Commonly due to penetration or rupture of the arterial wall following trauma (iatrogenic pseudoaneurysm).* The sac wall is lined by periarterial fibrous tissue, not by the layers of arterial wall.

Effects of aneurysm

Remember the mnemonics PTI (Press Trust of India)

- P Pressure
- T Thrombosis
- I Ischaemia

I. Pressure on the neighbouring structures

1. Pressure on the adjacent veins – can block the vein or cause thrombosis leading to congestion and oedema of the distal part e.g., popliteal aneurysm pressing over the popliteal vein leading to oedema leg.
2. Pressure on the adjacent nerves – causing altered sensation like pain, tingling and numbness, paraesthesia or rarely paralysis.
An aortic arch aneurysm may give pressure on the recurrent laryngeal nerve causing aphonia.
3. Pressure on the adjacent bones – causing erosion of the bones e.g., a vertebral body, sternum, or rib giving rise to backache and local bony tenderness. The intervertebral discs are resilient structures and do not undergo erosion.
4. Pressure on the adjacent organ e.g.:
 - (i) pressure of an aortic aneurysm on the oesophagus causing dysphagia;
 - (ii) stomach may be pushed forward by an aneurysm of the abdominal aorta; ultimately the aneurysm may leak or burst into the stomach causing severe haematemesis.

5. Pressure on the adjacent skin: The aneurysm with increased size may bulge under the overlying skin; the overlying skin becomes stretched, red, oedematous and appears inflamed simulating an abscess. (*So, one must be very careful to exclude aneurysm if an abscess lies in the line of a known artery. One should aspirate first before making an incision, particularly when situated in the chest wall, axilla, groin or popliteal fossa.*)

II. Thrombosis

There may be formation of a classical laminated thrombus, consisting of alternate layers of pale platelet thrombus and red blood clot. The process is slow, but eventually the sac becomes filled with thrombus. It may be protective by preventing rupture but it often causes ischaemia in the distal territory. This is a common occurrence in popliteal aneurysm.

III. Ischaemia

This may be due to one of the following causes:

- (i) due to pressure of the aneurysm on nearby branches of the artery;
- (ii) due to occlusion of the ostia of emerging vessels by the contained thrombus;
- (iii) due to embolism from contained thrombus, e.g., formation of multiple small emboli from subclavian aneurysm which block the digital arteries leading to gangrene.
- (iv) due to dissection of the vessel wall.

Complications and terminations

1. *Pressure on the adjacent structures as mentioned under 'Effects'.*
2. *Circulatory insufficiency* due to *thrombosis* or *embolism* may lead to ischaemia and gangrene of the distal territory.
3. *Rupture* → *Haemorrhage* – may be fatal. Rupture frequently occurs due to avascularity of the vessel wall; more common at the sides of the sac. The blood extravasates into the surrounding tissues (retroperitoneal haemorrhage, sub-arachnoid or intracerebral haemorrhage), or the sac may rupture in the peritoneal or pleural cavity (haemoperitoneum or haemothorax), in the oesophagus, stomach or duodenum (causing haematemesis). During rupture the patient may complain of severe pain associated with severe shock. The sudden onset of a severe headache in a middle-aged hypertensive patient is suggestive of spontaneous intracranial haemorrhage due to rupture of Berry aneurysm.
There may be double vision due to 3rd nerve palsy. This is caused by compression of the 3rd nerve just before entering the cavernous sinus by the expanding aneurysm or the adjacent haematoma.
4. *Infection* – usually arises from the organisms in the blood stream. Signs of inflammation may supervene, and if untreated, suppuration, abscess and rupture follow.
5. *Spontaneous cure* – may be brought about by the formation of clot and fibrosis within the aneurysmal sac leading to consolidation. This sometimes occurs in small peripheral saccular aneurysm.

Diagnostic criteria

They depend largely on the position of the aneurysm and its rate of growth.

Symptoms

1. Asymptomatic: When the swelling is detected accidentally during physical examination.
2. Symptomatic: Symptoms due to local cause when a patient may notice a *pulsatile swelling*. This is common with femoral or popliteal aneurysms – not so common with abdominal aorta aneurysm.

The patient may present with a dull aching pain with abdominal aneurysm; this dull ache is felt over the swelling in the centre of abdomen.

The aching pain is caused by stretching of the arterial wall. It does not radiate. It is not relieved or exacerbated by any natural event.

Acute pain can occur if the sac suddenly stretches or begins to tear.

A *severe pain, bursting in nature*, is common when the aneurysm suddenly ruptures resulting in a large haematoma.

Referred pain due to pressure on the adjacent nerve is not uncommon leading to pain and paraesthesia.

Patient may present with the symptoms arising due to the pressure effects on the neighbouring structures by the aneurysm – see effects of aneurysm (p. 246).

Aortic dissection produces intense pain, often described as tearing or shearing knife like. It is sudden and very severe. The pain of ascending aorta dissection is felt in the anterior chest wall and that of descending aorta dissection is felt posteriorly between the scapulae. The pain can migrate downwards into the flank or pelvis as the dissection process propagates distally.

Signs

May be intrinsic or extrinsic.

A. Intrinsic:

1. *Pulsatile swelling* in the course of an artery – both visible and palpable.
2. Swelling exhibits *expansile pulsation* and this is synchronous with the heart beat.
3. *On palpation – thrill may be palpable*.

4. Swelling is *cystic and compressible* – direct compression may empty the sac or diminish its size.
5. *On auscultation – systolic bruit* may be detected.
6. *Pressure proximal to the swelling* – the swelling diminishes in size and also the pulsation diminishes or ceases. On release of pressure there is reappearance of the swelling with expansile impulse.
7. *Pressure distal to the swelling* – the swelling increases in size, so also the expansile impulse.

B. Extrinsic:

1. Distal pulsation on the side of aneurysm found to be delayed and diminished, when compared with the pulse in the corresponding vessel of the opposite side.
2. Signs due to pressure effects on the adjacent structures:
 - (i) pressure on the veins: venous congestion with oedema of the leg e.g., in popliteal aneurysm;
 - (ii) gangrene;
 - (iii) pressure on the nerves: referred pain, paraesthesia or even paralysis, e.g., foot drop in popliteal aneurysm, aphonia due to recurrent laryngeal nerve palsy in aneurysm of the arch of the aorta.
 - (iv) pressure on the neighbouring structures e.g.:
 - (a) pressure on the oesophagus by aortic aneurysm – causing dysphagia;
 - (b) pressure on the bones causing erosion (as evident by radiology) leading to backache and local bony tenderness when vertebra is involved.

Differential diagnosis

1. *Swelling under an artery*: An artery may be pushed forward by an underlying structure and thus rendered prominent e.g., the subclavian artery by a cervical rib.
2. *Swelling over an artery*: Transmitted pulsation is liable to be mistaken with the expansile pulsation. Careful examination and postural change will differentiate it from expansile pulsation e.g., a pseudopancreatic cyst lying in front of the abdominal aorta will show a pulsatile swelling in the epigastric region. A pseudo-

pancreatic cyst, when examined in the genu-pectoral position, falls away from the aorta, and consequently pulsation is less definite or absent.

3. *Pulsating tumours* e.g., aneurysmal bone cyst, bone sarcoma, osteoclastoma, and a metastasis especially from a hypernephroma. Careful radiological investigation will differentiate these conditions from aneurysm.
4. *An abscess*: Already mentioned under Complication.
5. Arteriovenous fistula.

Differentiation between expansile pulsation and transmitted pulsation

Expansile pulsation

An aneurysm is characterized by expansile pulsation. When the two fingers are placed over the sides of a pulsatile swelling, in case of expansile pulsating swelling the two fingers are not only lifted up but are also separated from each other e.g., popliteal aneurysm.

Transmitted pulsation

When the two fingers are placed over the sides of a pulsatile swelling situated over a vessel, the two fingers are lifted up but are not separated from each other e.g.,

- (i) Cervical rib pushing the subclavian artery up giving rise to pulsatile swelling in the neck;
- (ii) Pseudopancreatic cyst may feel like a pulsatile swelling in supine position, as the swelling is situated in front of abdominal aorta. On postural change (knee elbow position) the swelling will be shifted away from the artery and thus will lose pulsation (cf. aortic aneurysm).

Investigations

1. Blood for lipids – Atherosclerosis.
Blood for W.R. and Kahn to exclude syphilis.
2. Radiology:
 - (a) Straight X-ray may reveal calcification of the arterial wall, soft tissue shadow or bony erosion.
 - (b) Arteriography to study the collaterals above and below the lesion.
Arteriography may indicate the scope and nature of operation required. But it is not of

much value in the diagnosis, because, due to the pressure of laminated mural blood clot, the aneurysmal dilatation is seldom visualised.

3. CT or MRI shows the 3-dimensional structure of the aorta.
4. Ultrasound.
5. Transoesophageal echocardiogram (TEE).
6. *Duplex ultrasound scanning is more precise test*:
 - (a) for determining the flow patterns and imaging of both venous system and arterial system;
 - (b) for providing accurate localisation of fistula site in an arteriovenous fistula.
7. MRI angiogram.
8. Ophthalmoscopy – to ascertain the degrees of atherosclerotic changes in the retinal vessels.
9. Renal function tests – Blood urea, serum creatinine, I.V.P. and isotopic renogram. These are necessary before undertaking any major surgery for abdominal aneurysm.

Treatment

There are various operative procedures which can be remembered by the mnemonics 'WEAL'.

- | | | |
|---|--|---|
| W | 1. Wiring
2. Wrapping | } <i>Reinforcing procedures</i> |
| E | 3. Excision only
4. Excision and end-to-end
5. Excision and grafting
6. Exclusion and bypass grafting | |
| A | 7. Aneurysmorrhaphy | < <i>Reconstructive</i>
< <i>Obliterative</i> |
| | 8. Amputation | |
| L | 9. Ligation of the artery | < <i>Proximal</i>
< <i>Distal</i>
< <i>Both</i> |

Aim of operation

- (i) To prevent the onset of complications.
- (ii) To preserve the blood supply to the distal territory.

Choice of operation

This depends upon—

- (i) the site of the aneurysm;
- (ii) the size of the artery concerned;
- (iii) the state of the collateral circulation.

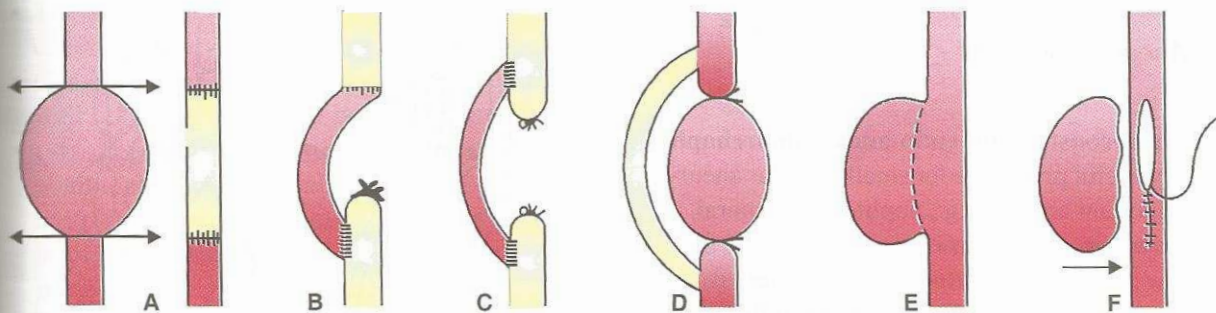


Fig. 15.3. Different methods of operation for aneurysm. A, Excision and end to end graft; B, Excision and end to side graft; C, Excision and side to side graft; D, Exclusion and bypass grafting; E & F, Matas aneurysmorrhaphy.

In most of the larger arteries aneurysm should ideally be treated by excision combined with some reconstructive procedures designed to maintain the patency of the artery.

In the cases of peripheral aneurysms, the nutrition to the distal territory is usually maintained by the collateral channels. So, the aim of operation here is to reduce or abolish the flow of blood through the aneurysm, if necessary by its *obliteration* or *excision*.

The treatment of aneurysms by proximal and/or distal ligation, intrasaccular wiring, wrapping in cellophane or synthetic cloth, and aneurysmorrhaphy is seldom used now-a-days because of the recent advances in the reconstructive vascular surgery technique and the advent of synthetic graft.

1. Wiring of the aneurysmal sac

It is indicated in elderly and poor risk patients with a difficult aneurysm so situated that operative excision carries an unduly high mortality.

A long fine thread of stainless steel wire (No. 3 and 200 to 1000 ft. in length) is introduced inside the aneurysmal sac with the help of a hypodermic needle. The wire gets coiled inside the sac and cause clotting of blood within the sac except a central channel which remains open. The cure is expected by clotting followed by fibrosis. Both the wire and the thrombus provide a strengthening framework for the weak aneurysmal sac.

2. Wrapping of the aneurysmal sac

The aneurysmal sac is wrapped by either a strip of fascia lata, or synthetic cloth, or plastic material like polythene, cellophane sheet – the idea being reinforcement of the wall and thus preventing rupture.

This method may be of palliative value where surgery is impossible e.g., intracranial aneurysm.

3. Excision only

Small peripheral aneurysms can be excised without endangering the peripheral circulation, e.g. aneurysm of the radial artery or the arteries which make up the circle of Willis.

4. Excision and end-to-end anastomosis

Small peripheral aneurysm is excised. The artery proximal and distal to aneurysm is mobilised and end-to-end anastomosis is performed.

5. Excision and grafting (Fig. 15.3A, B, C)

This is the ideal method and should be performed wherever possible. It consists of any one of the following:

- Excision of the sac and 'End-to-End' graft interposition (Fig. 15.3A).
- Excision of the sac and 'End-to-Side' graft interposition (Fig. 15.3B).
- Excision of the sac and 'Side-to-Side' graft interposition (Fig. 15.3C).

6. Exclusion and bypass grafting (Fig. 15.3D)

This method avoids damage to the adjacent vital structures (e.g., the popliteal vein in popliteal aneurysm, the vena cava in aneurysm of abdominal aorta). Here, the sac is ligated both above and below; the blood flow is redirected into the distal segment with the help of a bypass graft (Fig. 15.3D). The excluded aneurysmal sac becomes thrombosed, and rapidly shrinks into a fibrous mass, containing porridge like atheroma and blood clot in the centre. The by-pass graft used is mostly autogenous vein (e.g., reversed saphenous vein).

7. Aneurysmorrhaphy

- Reconstructive
- Obliterative

Matas reconstructive endo-aneurysmorrhaphy is a very useful procedure for small saccular aneurysm of important arteries e.g., popliteal or femoral.

Procedure: The sac is opened. Clots are removed. A new lumen is reconstructed for the artery from the sac wall (Fig. 15.3E & F) by suturing of the adjacent healthy arterial wall.

Obliterative endo-aneurysmorrhaphy can be carried out in traumatic or false aneurysm.

Procedure: The sac is opened. Clots are removed. The mouths of the feeding vessels are closed by ligature from within the sac. The sac is *obliterated* by further sutures of the sac wall.

Advantages:

- (i) Simple and safe method.
- (ii) Surrounding structures are safeguarded.
- (iii) Surrounding collaterals are left undisturbed.
- (iv) Effective in preventing recurrence.

8. Amputations

May sometimes be indicated in cases of aneurysm of the limb vessels with threatened or obvious gangrenous changes.

9. Ligation of the artery

This operation is becoming obsolete now-a-days. Simple ligature is one of the oldest operations in surgery. John Hunter's name is associated with ligation of the femoral artery in subsartorial (Hunter's) canal for the treatment of popliteal aneurysm.

The ligature may be applied either proximal or distal to the sac, or both, with or without intervention of the collaterals.

The idea was to reduce or abolish the blood flow within the aneurysmal sac by diverting the blood into the collaterals and thereby to induce thrombosis within the sac with subsequent fibrosis and shrinking and hence ultimate obliteration of the sac.

But the result is not always satisfactory, as complete obliteration is rarely achieved due to the return of blood within the sac either through the collaterals entering into it, or by the retrograde flow along the branches of the main vessels. In addition there is a

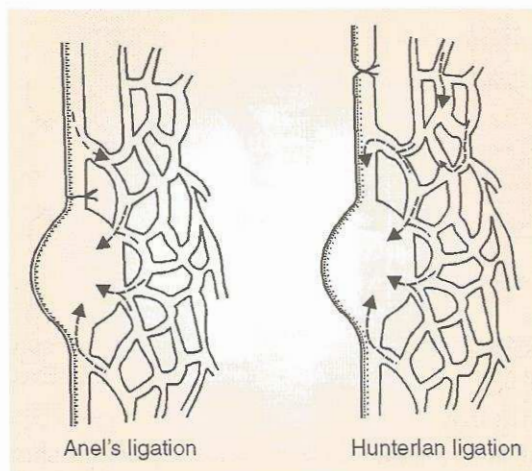


Fig. 15.4. Different methods of proximal ligation of aneurysmal sac. In either case blood may continue to return to the sac through the collaterals entering into it, or by retrograde flow.

great risk of gangrene unless there is adequate collateral circulation.

Site of ligation:

Proximal ligation (Fig. 15.4)

- Anel's ligation: The ligature is applied close to the aneurysmal sac.
- Hunterian ligation: The ligature is applied at some distance proximally.

Distal ligation

- Brasdor's ligation: The ligature is applied close to the aneurysmal sac.
- Wardrop's ligation: The ligature is applied at some distance distally.

Choice of site for ligation:

Aneurysms of the limb vessels may be treated by proximal ligation or by a combination of both proximal and distal ligation, with or without a sympathetic ganglionectomy. Proximal ligation may also be used in the treatment of intracranial aneurysms.

Aneurysms of the great vessels at the root of the neck are usually treated by distal ligation.

FALSE OR PSEUDO-ANEURYSM

- A false or pseudo-aneurysm is a sac formed by the condensed periarterial fibrous tissues.

- The sac communicates with the lumen of the contained artery through an opening in its wall.
- Commonly due to penetration or rupture of arterial wall following trauma, or iatrogenic injury.
- Iatrogenic pseudo-aneurysm occurs most commonly after arterial puncture for vascular access e.g. for aortogram or haemodialysis. *The most frequent site is the common femoral artery just below the groin.*
- It may also result from dehiscence of a surgical vascular anastomotic suture line – *anastomotic pseudoaneurysm.*
- Infection is a common disastrous problem and should be well taken care of.
- Pseudoaneurysm manifests with pain, a pulsatile mass or effects of compression in the adjacent structures.
- Duplex ultrasonogram may be helpful.

Treatment of false/pseudo aneurysm

Management includes one of the followings:

- Pseudo-aneurysm less than 2 cm in diameter have a 70% likelihood of spontaneous thrombosis with compression therapy.
- Larger one requires surgical interference.
- Pseudo-aneurysm arising in small, non-vital arteries may be treated with ligation, compression or coil embolisation.
- Others require direct surgical repair or exclusion of aneurysm with a stent coil.
- Anastomotic pseudo-aneurysm requires surgical repair and consists of patching or graft replacement of the disrupted segment.

ARTERIOVENOUS ANEURYSM OR FISTULA

This is a condition in which an abnormal communication between an artery and a vein is established. It may be either a *congenital* malformation, or *acquired due to the trauma* of a penetrating wound or a sharp blow.

The communication may be direct, when it is known as Aneurysmal varix, or by means of an intermediate sac between artery and vein, when it is called Varicose aneurysm (Fig. 15.4).

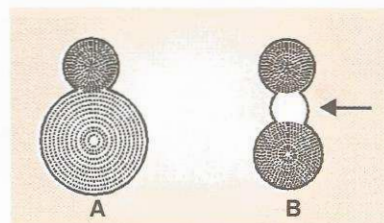


Fig. 15.5. Arteriovenous fistula. A, Aneurysmal varix; B, Varicose aneurysm.

Effect

Structural effect: As the arterial blood directly drains into the adjacent vein, the vein becomes gradually dilated, tortuous and thick-walled (arterialised). The collaterals also increase in size and become tortuous to maintain the peripheral circulation and thus make the lesions diffuse. Hence operation is difficult and hazardous and this is more so with the congenital variety.

Physiological effect: Due to diversion of blood from the normal arterial bed, there is a marked fall of diastolic pressure with a corresponding rise in the pulse pressure.

Increased venous return to the heart will result in an increase in pulse rate and cardiac output. Ultimately, left ventricular enlargement and later, cardiac failure will occur. So operation is indicated before cardiac decompensation has become established.

Types

1. Localised
2. Diffuse

In the diffuse congenital variety, which is relatively common, the arteriovenous communications are deep (may be in the bone) and clinically indiscernible.

Diagnostic criteria

Features suggestive of arteriovenous fistula formation vary according to whether it is localised or diffuse, congenital or traumatic.

Local signs

1. When the communication is superficial in situation, the patient may come with a *pulsatile swelling in a limb.*
2. *Presence of distended superficial veins* which

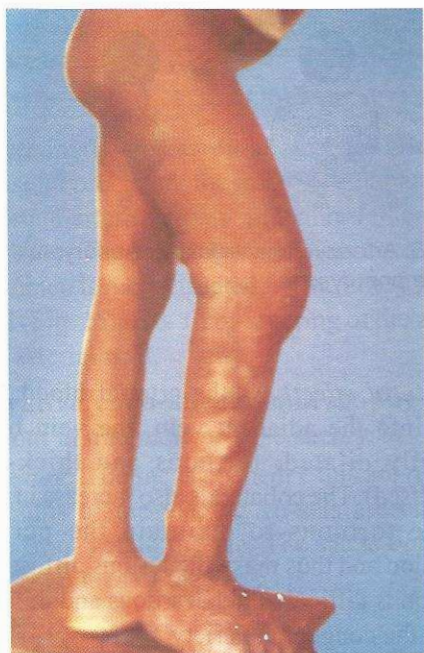


Fig. 15.6. Gigantism of the right lower limb in an 8-year-old child. This was due to arteriovenous fistula. Note the varicosities of the veins.

may be visible even when the patient lies down. Ultimately, the veins become *varicose*.

3. If there is presence of *Portwine discoloration of the skin* in addition – then the combination is highly suggestive of arteriovenous fistula.
4. Presence of superficial angiomas suggestive of congenital fistula.
5. The patient may present with *leg ulcer* (which is due to diversion of blood away from the skin). Often the ulcer is extremely painful. The ulcer is also known as hot ulcer as the surrounding skin feels warmer than normal.
6. The patient may present with *gigantism of the limb* i.e., increased growth of the limb supplied by the affected vessel. This is common in congenital fistula. It may be acquired, if the fistula occurs before the completion of epiphyseal union of the long bones.
7. Locally the extremity is appreciably warmer and usually moister than that of the unaffected side. Below the fistula the limb will feel cooler.
8. Muscle wasting below the fistula.
9. *On palpation – a thrill*, and *on auscultation – a buzzing, continuous bruit* (machinery murmur)

may be present, particularly in the traumatic type.

10. Pressure on the artery proximal to the fistula causes the swelling to diminish in size and the thrill and bruit to cease.

Systemic signs

1. *Branham's sign*: When the fistula-shunt is large enough, digital occlusion of the feeding artery proximal to the fistula causes slowing of the pulse rate and rising of the blood pressure. (The slowing of the pulse rate is due to vagus reflex effect for the sudden rise of vascular peripheral resistance. The sign becomes negative, if the vagus is blocked by atropine.)
2. Pulse-pressure is elevated.
3. In advanced state, left ventricular enlargement and, later, cardiac failure will be evident.

Investigations

1. Arteriography: This is a valuable test to know the site of fistula and also to find out the speed of venous filling.
2. Colour duplex ultrasonography may be helpful.
3. MRI angiogram may be helpful.

Treatment

1. Fistula affecting the smaller vessels: *Quadruple ligation* is the method of choice. Both the involved artery and the vein are ligated both above and below. Ligatures are also applied to any other vessels communicating with the sac. Thereafter, the sac may be obliterated with further ligatures. Every effort is made to avoid damage to important collateral vessels, on which the circulation of the limb depends.
2. Fistula affecting the large vessels: *Reconstructive operations are indicated*:
 - (i) The affected segment of the artery is excised and the gap is replaced by an autogenous vein graft or by a plastic prosthesis.
 - (ii) Occasionally, the artery can be reconstructed at the expense of the vein.
3. Simple ligation of the feeding artery alone proximal to the fistula, is contraindicated because of a chance of gangrene. (The reason for this is that the collateral circulation lacks the force to drive the blood into

the capillary bed distal to the fistula; it takes instead, the course of least resistance through the fistula back to the heart).

4. Selective intra-arterial embolisation is tried in some centres with success.
5. Stapling of the epiphyses may be attempted to prevent to rapid a growth of the limb.
6. Amputations may occasionally be required in the following conditions:
 - (a) when the giant limb incapacitates the patient;
 - (b) in case of intractable pain or gangrene;
 - (c) if the associated increased cardiac output leads to cardiac failure.

IATROGENIC ARTERIOVENOUS FISTULA OR SHUNT

When surgeons make an arteriovenous shunt for a

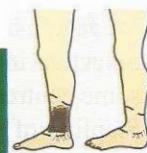
definite purpose as for haemodialysis. Commonly done at the wrist. The diagnosis can be assumed in case of:

- (i) Presence of pulsatile and expansile swelling in the vicinity of an accessible artery and a vein e.g., at the wrist or ankle.
- (ii) Presence of overlying scar mark.
- (iii) History of operation.
- (iv) Thrill on palpation.
- (v) Bruit on auscultation.

This type of temporary arteriovenous shunt or fistula is known as *Cimino fistula* and commonly noticed at the wrist using the radial artery and the cephalic vein, or at the ankle using the posterior tibial artery and a nearby vein.

16

CHAPTER



Varicose Veins

Varicose veins may be described as elongated, dilated and tortuous veins. The varicose veins bulge outwards under the skin.

Common sites of varicosity

1. Superficial venous system of the lower limb.
 - (a) Long saphenous vein.
 - (b) Short saphenous vein.
2. Haemorrhoidal venous plexus.
3. Oesophageal varices – involving the veins at the oesophagogastric junction and bulge inwards under the mucous membrane – visible only with upper GI endoscopy.
4. Varicosity of the pampiniform venous plexus (varicocele).

Only the varicosity of the saphenous venous system will be discussed in this chapter.

VARICOSITY OF THE SAPHENOUS VENOUS SYSTEM

SURGICAL ANATOMY

Venous system of the leg can be considered under three headings:

1. Superficial
2. Deep
3. Perforating

Superficial venous system (Figs. 16.1 and 16.2)

Comprises the long and short saphenous veins and their tributaries. They have the following characteristics:

- (i) They lie in the superficial fascia between the skin and the deep fascia, being closer to the skin, so often are visible when engorged. *Saphenous means easily seen.*
- (ii) They are low pressure and poorly supported system.
- (iii) The middle coat of these vessels is much thicker than that of the other veins and consists mostly of smooth muscles with little fibrous and elastic tissue.
- (iv) They are provided with numerous valves.
- (v) Normally the blood in the superficial venous system flows into the deep venous system, except in the sole (and also in the palm) where blood in the deep venous system flows into the superficial venous system.

The **long saphenous vein (LSV)** is the longest vein of the body. It commences from the medial side of the dorsal venous arch of the foot and courses in front of the medial malleolus and ascends to the posteromedial side of the knee. Then the vein inclines laterally and forward in the thigh towards the saphenous opening which lies $1\frac{1}{2}$ inches (3.8 cm) below and lateral to the

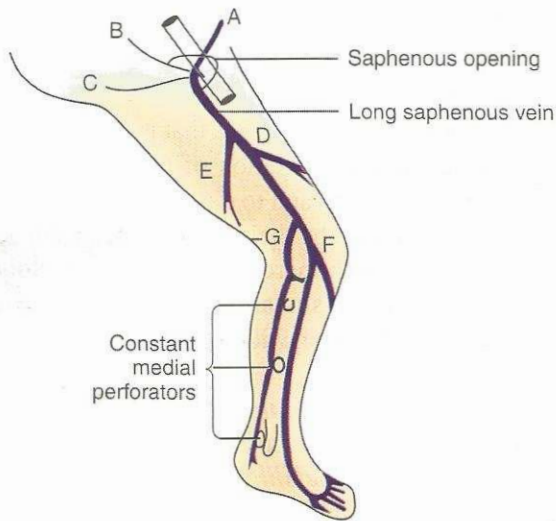


Fig. 16.1. Long saphenous vein and its tributaries. A, Superficial external pudendal vein; B, Superficial epigastric vein; C, Superficial circumflex iliac vein; D, Anterolateral superficial femoral vein; E, Postero-medial superficial femoral vein; F, Anterior vein of the leg; G, Posterior arch vein which lies parallel to and behind the main trunk of long saphenous vein. It anastomoses with the small venous arches connecting the medial perforating veins (H).

pubic tubercle. It pierces the cribriform fascia and passes deeply to enter the femoral vein. The vein contains more than 12 valves. *The saphenous nerve accompanies the vein in the leg.*

Tributaries of long saphenous vein are shown in Fig. 16.1.

The **short saphenous vein** commences from the lateral side of the dorsal venous arch and passes behind the lateral malleolus and then ascends obliquely backward towards the middle of the popliteal fossa, where it pierces the deep fascia to enter the popliteal vein in most cases. *The sural nerve accompanies this vein in the lower third of the leg.*

Perforator system

The perforating veins are communicating veins between the superficial and deep veins. They perforate through the deep fascia and connect the two venous systems. They are also provided with the valves near their origin and at their entrance to the deep veins. *Under normal conditions they allow blood to flow only from the superficial to the deep*

veins. Reversal of blood flow occurs due to incompetence of perforators which will lead to varicose veins. This system comprises following groups:

1. **Medial perforating veins** (Fig. 16.1): These are three constant medial leg perforators and *situated along the posterior border of the tibia* 2 inches (5 cm), 4 inches (10 cm), and 6 inches (15 cm) successively above the medial malleolus. *The upper two of these perforators enter the posterior tibial vein at the precise level where one of the unvalved 'soleal venous sinuses' enters* (Fig. 16.3). So, the clot arising in the soleal veins may extend into the posterior tibial vein and then into these perforating veins, thus destroying the mechanism which obviates reflux. All these medial perforators again are connected by way of the posterior arch vein to the long saphenous vein (Fig. 16.1G).
2. **Lateral perforating veins** (Fig. 16.2): These are inconstant in their positions and *situated along the posterior border of the fibula* 2 inches (5 cm), 5 inches (12 cm), and 7 inches (17 cm) above the lateral malleolus and are connected with the peroneal veins or venae comitantes.
3. **Central perforating veins**: Usually one or two veins connecting the short saphenous system to gastrocnemius and soleus muscles. The former enters its muscle on the medial side close to its junction with the tendoachillis while the latter, further up the calf.

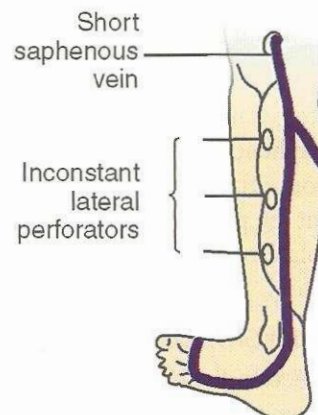


Fig. 16.2. Short saphenous vein and inconstant perforating veins.

Deep venous system (Fig. 16.3)

This is a high pressure system. This accompanies the main arteries and is well supported by powerful leg muscles which compress the veins. It is the powerful muscle contraction that helps in return of the blood to the heart. *The deep vein is also provided with unidirectional valves.*

This deep venous system comprises:

- (i) Femoral and popliteal vein
- (ii) Venae comitantes (paired veins) accompanying the anterior tibial, posterior tibial and peroneal arteries.
- (iii) Soleal venous sinuses. Valveless veins draining the calf muscles.

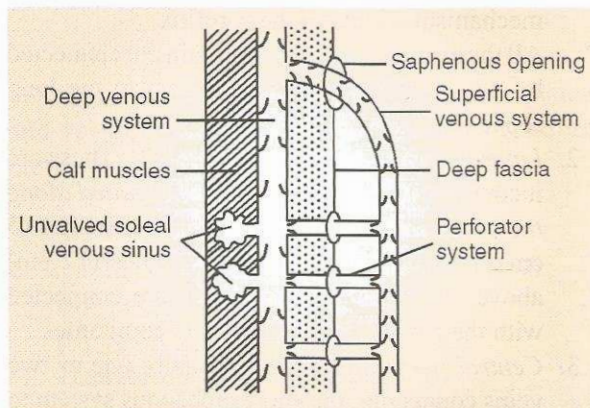


Fig. 16.3. Disposition of superficial and deep venous systems of the lower limb with inter-communication between them.

This deep venous system communicates with the low pressure superficial venous system at the following sites:

- (i) Saphenofemoral junction – constant.
- (ii) Adductor canal by inconstant perforators.
- (iii) Short saphenopopliteal junction – constant.
- (iv) Posteromedial aspect of the leg by three constant leg perforators.
- (v) Posterolateral aspect of the leg by inconstant perforators.

SURGICAL PHYSIOLOGY

Blood is returned to the heart from the lower limbs by the following mechanisms:

1. *Vis-a-tergo* of the circulation, that is, the pressure transmitted from the arterial tree passes through

the capillary bed to the venous side. Pressure in the arteriolar end of the capillary is 32 mm Hg and in the venular end of the capillary is 12 mm Hg.

2. *Calf-pump or muscle pump*, that is, the alternate contraction and relaxation of the surrounding muscles of the leg and to a certain extent of the thigh surrounded by their firm fascial sheath act as a pump which increases the return of blood to the heart. A sustained contraction or a continued relaxation will not operate the muscle pump.
3. *Competent valves in the veins* – normally the veins of the limbs are provided with *unidirectional competent healthy valves* which allow the flow of blood in one direction, that is, towards the heart. These valves break up the column of blood and thus prevent distension of the veins due to gravity.
4. *Venae comitantes* – These lie by the side of the arteries and may be helped by the arterial pulsation to propel the blood upwards to the heart.
5. Negative intrathoracic pressure during inspiration is transmitted to great veins and causes drop in venous pressure which aids in venous return.

In the resting position blood returns to the heart mainly by a vis-a-tergo and *during walking* mainly by the calf-pump. In addition, the actual pressure in any one vessel is also dependent on the effect of gravity. On standing, therefore, the actual pressure in any vein includes a hydrostatic pressure (up to pressure of 90 mm of Hg) which is equivalent to the weight of the column of blood from the legs to the heart, and this pressure is exerted on the valves, particularly on those guarding the communications between the superficial and deep venous systems of the leg.

The above factors will be effective only if the valves are competent enough to support the column of blood passing *each valve 'check point'* and the gravitational effects are reduced to a minimum.

Development of varicose veins

Varicose veins develop when the blood leaks continuously from high pressure deep venous system and is allowed to enter the low pressure and poorly supported superficial venous system. Under normal

conditions the blood in the superficial venous system flows into the deep veins and the superficial system is protected from reflux of blood from the high pressure deep venous systems by the *unidirectional strategic valves in the deep and perforator venous systems*. But if this mechanism breaks down, there will be retrograde flow of blood in the superficial veins leading to distension and tortuosity.

Aetiology

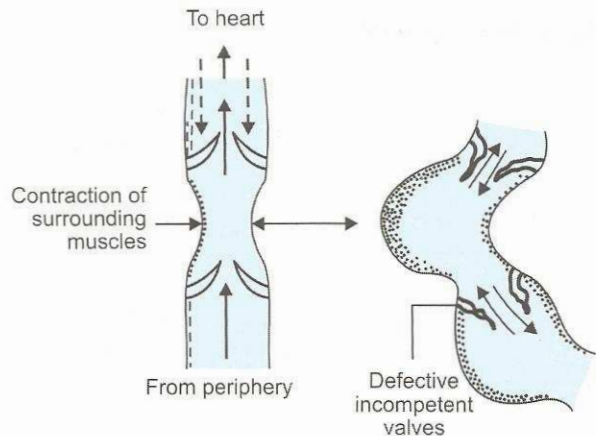
Can be considered under two headings – primary and secondary

1. Primary or idiopathic

- Exact cause is not known.
- Often familial with onset during young adulthood or pregnancy.
- *Commonly due to weakness of the venous wall that permits valve ring dilatation or congenital incompetence or absence of the valves.*
- **Exciting causes:**
 - (a) *Prolonged standing and/or sitting* in one position whereby the gravitational pressure of a long column of blood presses continually upon a weakened valve. It also deprives the veins of the benefits of muscular pump movement.
 - (b) *Violent muscular efforts* which are responsible for the frequency of varicose veins in the legs of athletes. It is postulated that excessive continuous calf muscle activity may actually force the blood through the perforating system in a reverse direction and *thus causing perforator valve disruption*. This factor is well illustrated by the fact that in our country more commonly Rickshaw-pullers suffer from varicosity of vein.
 - (c) *Obesity*: Soft, virtually fluid, fatty tissue offers poor support to the veins.
 - (d) *Ageing* – causes atrophy and weakness of the vein wall.

2. Secondary

- *Obstruction to venous outflow* due to following reasons:
 - (a) Pregnancy
 - (b) Chronic constipation with loaded colon
 - (c) Abdominal or pelvic tumour like uterine fibroid, ovarian tumour or cyst or pelvic cancers.



Normally the veins are provided with unidirectional competent healthy valves – so, no backflow

Allows backflow

Fig. 16.4.

- (d) Retroperitoneal lymphadenopathy
 - (e) Retroperitoneal fibrosis
 - (f) Ascites
 - (g) Prior deep vein thrombosis (DVT).
- *Local damage to veins and valves by an inflammatory or traumatic process* leading to thrombophlebitis or phlebothrombosis. This causes obstruction of the venous flow. *Also it may destroy the valves*. Following recovery, after an attack of thrombosis, the veins subsequently recanalise but their valves are rendered weak and incompetent.
 - *Congenital or traumatic arteriovenous fistula* – the high pressure arterial blood directly drains into the adjacent vein through the abnormal communication thereby causing distension and varicosity of the veins.
 - *Extensive cavernous haemangioma*.
- These last two causes should be considered when varicosities are found in persons below the ages of twenty.*

Incidence

- Majority (60–70%) develop as a result of saphenofemoral junction (SFJ) incompetence and long saphenous vein (LSV) reflux.
- 10% develop due to saphenopopliteal junction (SPJ) incompetence and short saphenous vein (SSV) reflux.
- Remainder arise due to both SFJ incompetence and perforator incompetence.

Diagnostic criteria

1. **Age:** Varicose veins affect all age groups but common in young and middle-aged.
A varicose vein in children is invariably due to congenital vascular abnormality.
2. **Sex:** Equally occurs in both sexes. But women are more likely to present early for treatment because of cosmetic reasons.
3. **Occupation:** Many patients with varicose veins have jobs that involve prolonged standing or sitting e.g., traffic police, bus or tram conductors, shop assistants working in shopping malls. In our country rickshaw pullers also suffer from varicose veins.

Symptoms

1. *Tiredness, heaviness and dull aching sensation felt in the lower leg or in the calf*, particularly worse at the end of the day resulting after prolonged standing or sitting. The aching is due to pooling of blood and venous distension in the limbs. *The aching is relieved by elevation of the limb and exercises.*

Some patients suffer from *night cramp in the calf* shortly after retiring to bed. This is due to sudden change in the caliber of the communicating veins which stimulate the muscles between which they pass. *The cramp is also relieved by elevation of the limb or exercises.*

This aching pain or cramps should not be confused with those due to peripheral arterial disease referred to as *intermittent claudicating pain where the pain is made worse by exercise but that of varicosity is improved.*

The pain may be bursting and severe in nature with deep vein thrombosis, and may be localised at the site of perforating veins when perforator incompetence likely to be present.

2. The patient may present *because of cosmetic disfigurement* with multiple dilated and tortuous visible veins in the leg and/or with its complications such as:
 - (a) Swelling of the leg mostly round about the ankle joint.
 - (b) Venous stars, blowout or visible varix – a localised dilated segment of vein.
 - (c) Pigmentation.

- (d) Eczema – dry and scaly or moist, itchy and infected.
 - (e) Venous ulcers in the lower 1/3rd of the leg.
 - (f) Bottleneck deformity of the leg.
3. History of occupation which involves prolonged standing or sitting.
 4. Past history: The patient should be asked about any previous episodes of leg swelling, thrombophlebitis or white leg following operation, accident or pregnancy that may have produced damage to the valves of the perforating veins and/or the deep venous system.

Signs

1. Presence of dilated, tortuous visible veins in the leg and the thigh.
2. The skin, particularly on medial side of the lower leg (Gaiter area), is inspected for blowout, oedema, brawny induration, pigmentation, ankle flare, ulceration or healed scars.
 - *Blowout* (visible varix) – A localised dilated segment of vein. It signifies underlying perforator.
 - *Ankle flare* – Telangiectatic dilated reticular veins caused by the distension of superficial veins. This is seen around the ankle, most apparent over a triangle centered at the medial malleolus (varicular triangle).
 - *Healed scar* – Signifies previous ulceration.
3. **Saphena varix:** This is a saccular dilatation at the upper end of the long saphenous vein at the saphenous opening. The saphenous opening lies medial to the femoral artery below the inguinal ligament about 1½" below and lateral to be pubic tubercle.

Saphena varix has the following features:

- (a) It is compressible but non-pulsatile (cf. femoral aneurysm).
- (b) It has a positive cough impulse (cf. femoral hernia).
- (c) It disappears when the patient lies down (cf. femoral hernia where spontaneous disappearance is unlikely, and requires manipulation).
- (d) The varix has a *fluid thrill* which can be felt over the lump in the groin only when the long saphenous vein lower down the thigh is tapped.

Clinical tests

There are many clinical tests for examining the varicose veins, which are necessary to diagnose as well as to determine the choice of treatment. For convenience they may be considered in three main groups:

- (a) to know which venous system is varicose.
- (b) to know which perforator or perforators are incompetent.
- (c) to know the patency of the deep veins.

Which venous system is varicose—long or short saphenous venous system?

Inspection: Both the front and back of the lower limb with the patient standing are inspected to determine the veins affected.

In affection of the long saphenous system, varicose veins are seen on the medial side of the leg and thigh proceeding up towards the saphenous opening at the groin.

In affection of the short saphenous system, varicose veins are seen on the back of the leg proceeding up towards the popliteal fossa.

Look for any local signs of complications of varicose veins (see under symptoms) especially on the medial side of the lower third of the leg.

Schwartz test: With the patient in standing position two fingers of the left hand are placed over or just below the saphenous opening (fossa ovalis) and with the right middle finger the most prominent part of the varicosity lower down the leg and/or thigh is tapped – a *thrill-like impulse* will be felt by the fingers at the saphenous opening particularly in presence of saphena varix. It signifies continuous column of blood due to valvular incompetence.

Which perforator or perforators are incompetent – saphenofemoral perforator or leg perforator?

I. Cough impulse test: To determine the saphenofemoral incompetence.

The fingers are placed over the course of the vein just below the saphenous opening and the patient is asked to cough – a palpable fluid thrill (and a bruit on auscultation) are evident, if the valve at the saphenofemoral junction is incompetent.

II. Trendelenburg test: To detect the incompetence of the saphenofemoral junction and/or the other communicating system.

The patient lies down on the couch in supine position, the limb is elevated above the level of the heart to allow the varicose veins to empty (which can also be helped by stroking the veins proximally). Then the thumb is placed firmly over the saphenous opening or an elastic rubber tourniquet is applied firmly round the thigh at the saphenous opening. Now keeping the firm pressure over the saphenous opening, the patient is asked to stand up quickly. *The findings and the remarks are:*

- (i) The pressure is not released but maintained for about one minute. If there is gradual filling of the veins from below during this time – it indicates incompetence of the leg perforating veins allowing retrograde flow.
- (ii) The pressure is released. If there is quick filling of the veins by a column of blood from above – it indicates incompetence of the valves at the saphenofemoral junction.

Both the above groups (Nos. i and ii) are regarded as Trendelenburg test positive and are indications for operation.

If the short saphenous system is involved a similar test can be performed with the thumb or tourniquet pressure over the upper end of the vein at the level of knee crease.

III. Pratt's test: Test for leg perforators.

An elastic compression bandage is applied from toe to upper thigh which causes emptying of the varicose veins. A tourniquet is then tied around the upper thigh above the bandage. Now, with tourniquet in position, the bandage is unwinded in downward direction – a *'blowout'* i.e., a *visible varix* will appear over the site of constant leg perforators indicating the incompetence of the perforators.

Marking these sites with inks helps during operation to look for the perforator pits.

IV. Fegan's test: For seeking the sites of perforator.

First, the varicosities are marked with a skin pencil with the patient standing. The patient lies down. The affected limb is elevated above the heart level and the heel is supported against the examiner's upper chest. Now, one is to palpate along the line of the marked varicosities carefully for any gaps or pits in the deep fascia which transmit the perforators – they

are felt as circular openings with sharp edges and are marked with an 'X'. These 'X' marked positions are compared with the anatomical sites of the perforating veins.

This test may also be confirmed by Trendelenburg test to determine whether the superficial varicosities fill from the perforators marked by 'X'.

Leg perforator incompetence may also be evident from the following signs:

(i) *An ankle flare* – telangiectasia of reticular veins caused by the distension of the superficial veins and is seen around the ankle most apparent over a triangle centred at the medial malleolus (varicular triangle).

(ii) *A 'blow-out' or a visible varix* i.e., a localised dilated segment of the vein – situated over the site of constant medial perforators.

Tests for patency of the deep veins

Perthes's test: *To know whether the deep veins are patent or not.*

This test should be carried out if the patient complains of a bursting pain in the lower leg on standing or walking. A tourniquet is tied tightly round the upper part of the thigh below the saphenous opening while the patient stands with the dilated veins being full, tightly enough to prevent any reflux down the vein. The patient is then asked to walk quickly for about 5 to 10 minutes. *The findings and the remarks are:*

- (i) If the varicose vein shrinks or disappears – it indicates competence of the perforating veins and deep veins.
- (ii) If the varicose vein remains unchanged or becomes more distended – it indicates that the perforating veins and deep veins are blocked. There will also be reappearance of pain of which the patient complains.

In the absence of deep veins patency, the superficial venous system is the only vein to drain the blood from the leg – so, operation is contraindicated.

Other examinations

- (i) In addition to the above tests the abdomen should be palpated in every case to exclude pregnancy or pelvic tumour because sometimes a pregnant uterus or an intrapelvic tumour may be responsible for the development of varicosity (secondary).

There may be dilated, tortuous, collateral veins crossing the abdomen which may indicate Inferior Vena Cava thrombosis.

- (ii) *The peripheral arterial pulses should be palpated* to ensure that there is no accompanying arterial insufficiency. Venous and arterial disease of the lower limb may coexist particularly in elderly men.
- (iii) P/R – To exclude pelvic tumour.

Investigations

A careful history and physical examination are usually enough to diagnose the primary or idiopathic varicose veins.

Phlebography may be useful:

- (a) to determine the patency of the deep veins;
- (b) to localise the sites of perforators.

In the past, phlebography was carried out routinely in some clinics.

Doppler ultrasound may be accurate:

- (a) to determine the patency of the deep veins;
- (b) to determine the presence of clot in the deep veins. No sound is heard in presence of deep vein thrombosis.
- (c) to detect the presence of incompetent valves either in deep veins or in the communicating veins.

Duplex ultrasound scanning is more precise test:

- (a) for determining the flow patterns and imaging of both superficial and deep venous system and of the arterial system.
- (b) for providing accurate localisation of the sites of perforating veins and fistula site in an arteriovenous fistula.

In the absence of deep vein patency, operation for varicose vein is contraindicated. – Why?
Because the superficial varicose veins are the only veins to allow venous return to the heart.

Complications of varicose veins

1. Pigmentation
2. Oedema of the leg
3. Brawny induration
4. Ulceration
5. Eczema
6. Periostitis
7. Calcification

8. Thrombophlebitis
9. Haemorrhage
10. Deformities of the foot.

Pigmentation: A brown or dark brown discoloration is usually seen in the medial aspect of the lower 1/3rd of the leg, superior to medial malleolus (**Gaiter area**). This is due to deposition of haemosiderin pigment derived from the break down of RBC following subcutaneous extravasation of blood which has come out of the thin dilated veins resulting from venous stasis.

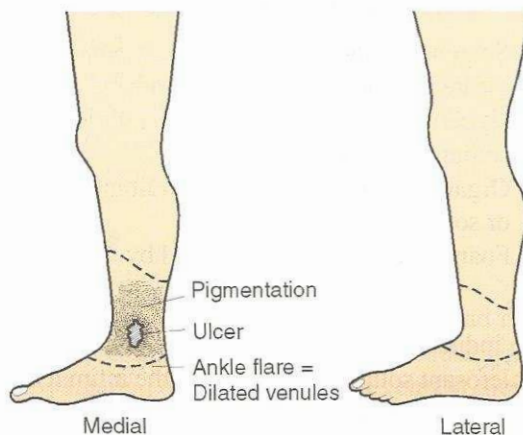


Fig. 16.5. Gaiter area of the leg.

Oedema of the leg: Soft and pitting. It may not be observed with isolated varicosity. It is usually present in patient with deep vein thrombosis and incompetent perforators.

Brawny induration: It is leg oedema but hard and indurated (feel like wood) and *non-pitting*. This is due to increase in the thickened connective tissue due to scarring secondary to fat necrosis, excess protein in tissue fluid, inflammatory changes and hyperemia leading to thickening of subcutaneous tissue giving a leathery texture to the skin. This is also referred as lipodermatosclerosis.

Eczema (chronic dermatitis): Patchy areas of eczema with or without itching may follow minor trauma or self-scratching of the skin, or an allergic manifestation resulting from local application of various ointments or strapping.

Ulceration: Venous stasis leading to increased venous pressure is the underlying cause as it results in local tissue anoxia leading to venous ulcers.

- More common in patients with deep vein abnormalities and/or incompetent leg perforators.
- Venous ulcers commonly develop over the medial aspects of the lower third of the leg, around and above the medial malleoli because of the presence of large number of perforators which transmit pressure changes directly into the superficial venous system. This area is called as *Gaiter's area*.
- The Gaiter's area is the common site for blowouts, ankle flare, induration, pigmentation and ulceration.
- Rarely, carcinomatous change may occur in chronic venous ulcer (*Marjolin's ulcer*).

Periostitis: Occurs in long standing cases if the ulcer is situated over the tibia.

Calcification: Occurs occasionally in the walls of the vein in long-standing varicosity.

Thrombophlebitis of the superficial vein: May occur spontaneously or may be secondary to trauma to the leg. It may be iatrogenic following injection of sclerosing fluid. It reveals itself as a reddened tender cord in the subcutaneous tissue. It may be recurrent and may extend in the deep venous system.

Haemorrhage: Usually due to minor trauma to the dilated veins. Such haemorrhage may occur into the subcutaneous tissue causing excessive bruising, or externally causing profuse bleeding due to the high pressure within the incompetent veins. *Bleeding generally stops if the limb is elevated and pressure bandage is applied locally. A tourniquet must not be used.*

Deformities of the foot: Talipes equinus due to the long continued faulty habit of walking on the toes to get relief of pain resulting in contraction and shortening of tendoachillis. Cases usually respond to remedial exercises.

Treatment

Patients with varicose veins seek treatment for one of the following reasons:

1. Severe aching or pain in the legs, particularly after prolonged standing.
2. Persistent or recurrent complications of varicose veins.
3. Because the leg is cosmetically unsightly.

- When varicose veins appear to be a bar to join into a particular job e.g., Nursing, Police or the Armed Forces.

Treatment of varicose veins

Three methods of treatment are available:

- Palliative treatment.
- Sclerotherapy.
- Operations.

Palliative treatment

Indications

- Aged.
- Pregnancy or puerperium.
- Patients who refuse operation or who are unfit for operation, or who are waiting for operation.
- Pelvic tumour.
- Deep vein thrombosis: confirmed by positive Perthe's test, venography or duplex ultrasonography.

Essentials

- Postural drainage:*
 - Avoidance of prolonged standing.
 - Elevation of the limb preferably above the heart level during rest or at night improves venous drainage and reduces any oedema.
- Exercises:* Exercises of the leg to strengthen the calf muscles which empty the veins are of value. 'Bicycle riding' in the air while lying on the back, walking and cycling are of obvious benefit.
- Support:* A crepe bandage or a well fitting 'two way stretch' elastic compression stocking, which includes the heel, should be applied before getting out of bed in the morning. It helps to compress the minor varicosities and to strengthen the efficiency of the calf-pump.
- Drugs used for varicose veins* particularly in the presence of oedema, pigmentation and ulceration are:
 - Calcium dobesilate – 500 mg BD. It improves both the venous and lymph flow and reduces oedema. Also improves macrophage-mediated proteolysis to help ulcer healing.
 - Dosmin 450 mg BD or Daflon 500 mg BD may be used in relieving night cramps. But they do not help in ulcer healing.

Sclerotherapy

Indications

- Small varicosities and cutaneous spider veins.
- Recurrent varicosities after operation.
- Varicose veins confined below the knee.
- Troublesome vulval varicosities.
- As an alternative to surgery.
- Cosmetic: But not ideal because of unpredictable staining which may occur.

Principle

Injection of a sclerosant solution into an empty varicose vein. The materials available are:

- Soap solution such as 5 percent ethanolamine oleate or 3 percent sodium tetradecyl sulphate.
- Hypertonic solutions of a crystalloid such as sodium chloride or glucose.
- Organic compounds such as quinine, urethane or sodium salicylate.
- Foam sclerotherapy developed by Tessari.

Idea

- To induce chemical thrombophlebitis.
- Sclerosant solution will damage the intimal lining of the vein so that an aseptic and firmly adherent thrombus, and later sclerosis develop leading to complete obliteration of the treated vein.

Method

The sclerosant solution should be injected into an empty varicose vein and *compression bandage* is then applied over the site which is maintained for a few weeks so that wall adheres to wall with no intervening blood clot; there will be formation of intravascular granulation tissue which later becomes fibrous and thus obliterates the lumen of the vein permanently.

Dose

The maximum dose at one sitting and at any one point is 1 ml. The injection can be repeated at various sites in the leg but the total dose should not exceed 10–12 ml.

When compression sclerosant therapy is used, a latex foam pad is applied over the site of injection and along the length of vein, which is maintained in position by crepe bandage and a full-length two-way stretch elastic stocking *for a period of six weeks* from the last injection or until the pain has gone from the site. *Compression bandage helps to prevent DVT.*

Sites of injections

1. At the maximum point of varicosity.
2. Sites of 'blow-out' or perforating veins.
3. At the maximum point of tenderness.

Complications due to injections

1. The injection should always be intravascular. Otherwise, skin ulceration may occur, which is more common with the crystalloids and the organic compounds.
2. Some risk of anaphylactic reactions.
3. Chance of deep vein thrombosis because of spread to the deep veins, if an excess of the sclerosant is injected at one site.
4. There will be local pain, oedema, redness, hyperpigmentation, etc., for a few days which will subside spontaneously.

Patient should be warned of these complications.

Operations

Aim of operation

1. To stop the retrograde venous flow (and hence high pressure leak) from the deep venous system, by ligation of the incompetent veins at the:
 - (i) Saphenofemoral junction
 - (ii) Saphenopopliteal junction and/or
 - (iii) At the incompetent perforator sites.
2. To remove the varicosities.

Method

- (i) Ligation alone.
- (ii) Ligation and stripping – The treatment of choice.
- (iii) Ligation and retrograde injections of sclerosing solutions to the distal segment.
- (iv) Multiple stab avulsion or excision of the convoluted veins.
- (v) Cockett and Dodd operation.

1. Long saphenous vein incompetence

The saphenofemoral junction is dissected out at the groin. All the tributaries are divided in between the ligatures. Then the saphenous vein is ligated flush with the femoral vein proximal to any tributaries, otherwise a recurrence is inevitable. This is known as **Trendelenburg operation**.

The Trendelenburg operation is nearly always supplemented with stripping of the varicose vein with

the aid of Myer's vein stripper so that the vein is avulsed from the subcutaneous tissue. Stripping of the varicose vein removes the unsightly veins and also disconnects the other tributaries of the main saphenous system.

After saphenofemoral flush ligation and disconnection, the long saphenous vein is exposed at the ankle in front of the medial malleolus, and a flexible wire vein stripper is introduced into the saphenous vein through a venotomy and pushed upwards towards the groin until it emerges from the proximal end of the vein. The vein is then fixed around the stripper at its lower end by a thread ligature and its distal end is divided and ligated. The ankle wound is closed. The stripper is then gradually pulled out through the groin incision while the leg is firmly bandaged with elastic crepe bandage by an assistant as the stripper advances.

Following stripping of the LSV, multiple stab avulsions of the localised dilated segments of the veins (blowouts) using mosquito forceps or avulsion hook can also be made.

Large tributaries may require separate exposure and ligation.

2. Short saphenous vein incompetence

The short saphenous vein is exposed by a curved transverse incision at the popliteal fossa and is ligated proximal to any tributaries close to or flush with the popliteal vein. *Stripping is usually not practised in short saphenous incompetence.*

3. Perforator vein incompetence

Incompetent perforator sites are explored subfascially and then ligated and divided. This is known as **Cockett and Dodd operation**.

In addition to above procedures, smaller varicose tributaries and severely dilated clumps of veins, if present, can be marked out before operation and may then be individually avulsed, ligated or injected by sclerosing solutions.

Contraindication of both injections and operative treatment

1. Women taking contraceptive pills, because there are chances of deep vein thrombosis and fatal thromboembolism. Pill alters the viscosity of blood.

2. Deep vein thrombosis with oedema of the limb because here the main axial deep vein has been destroyed and the superficial varicose veins form the major part of the venous drainage of the limb.
3. Pregnancy.
4. Thrombophlebitis.

Recent methods of minimally invasive therapy for varicose veins:

1. EVLA – Endo Venous Laser Ablation
2. RFA – Radio Frequency Ablation
3. SEPS – Subfascial Endoscopic Perforator Surgery

EVLA & RFA

Both cause thermal damage of endothelium resulting in endothelial denudation, collagen denaturation and subsequent fibrosis leading to venous occlusion.

Advantages:

- Both the methods can be performed in an outpatient department without anaesthesia.
- Patient can return to his normal activity early.
- Cosmetically good – because of no scars.
- Lower risk for recurrences.

Disadvantage: Cost is very high.

SEPS (Subfascial Endoscopic Perforator Surgery)

- Indicated for leg perforators.
- *Under laparoscopic guidance* subfascial perforators are identified and ligated.
- Procedure is simple and quick.
- Large incision is avoided.
- Morbidity is less.

17

CHAPTER



Ulcers of the Leg

The lower leg is the common seat of an ulcer and presents a common problem. 75 percent of cases are due to disorders of the venous system and the rest are due to a number of other possible causes including the 'Tropical ulcers' occurring in people living in the tropics.

ULCERS OF THE LEG

TYPES ACCORDING TO CAUSES

1. Venous
2. Arterial
3. Traumatic
4. Infective
 - (a) Specific
 - Tuberculous
 - Bairnsdale
 - Syphilitic
 - (b) Non-specific
 - Pyogenic
5. Neuropathic
6. Neoplastic
7. Cryopathic
8. Diabetic
9. Idiopathic
10. Self-inflicted or artefact
11. Miscellaneous: Ulcers of the leg may be associated with arteriovenous fistula, sickle cell

anaemia, chronic lymphoedema, various collagen disorders, poliomyelitis, ulcerative colitis, etc.

SURGICAL PATHOLOGY OF DIFFERENT VARIETIES OF ULCERS

Venous ulcer

Also called gravitational ulcer. *Always associated with incompetent venous systems* due to either varicose veins or deep venous thrombosis in which recanalisation of the deep vein has occurred, but the valves are either destroyed or incompetent due to damage. *The basic defect is venous stasis which results in local tissue anoxia due to stagnation of poorly oxygenated blood.*

As a result of incompetence of the valves of the deep veins and of the communicating veins between the deep and superficial venous systems, there is leakage of blood under high pressure from the deep veins into the superficial veins; this occurs particularly in the region of a constantly placed perforator over the medial side of the leg (the ulcer bearing area). This results in venular dilatation (ankle flare) and stagnation of blood. The blood in these varicose veins usually has only two-thirds of the normal oxygen content with a corresponding increase of carbon dioxide. There is also remarkable rise in pressure in varicose veins in the ankle, which may

be 30 mm higher than the blood pressure in the arteries particularly when the patient strains. Hence, the blood from the superficial capillaries is unable to enter the veins against this pressure, resulting in stagnation and congestion of the skin. The skin in this area is normally thin, delicate and easily traumatised. Stagnation of circulation and poor oxygenation combined with effused blood pigment *cause induration and pigmentation of the skin* which are the pre-ulceration changes. If the high pressure leakage is allowed to continue, skin ischaemia can occur and ulceration results, which may be precipitated by a minor trauma.

Arterial ulcer

The basic defect is skin ischaemia following atherosclerotic peripheral vascular disease. It commonly occurs on the toes, dorsum of the foot, or the heel and to start with there is a patch of dry gangrene.

Traumatic ulcer

Commonly occurs where the skin is closely applied to bony prominences, e.g., shin of the leg, malleoli and the back of the heel.

Plaster sores and bed-sores over the heel are included in this group.

Infective ulcer

(a) *Pyogenic*: Common organism is staphylococcus which may be so potent as to result in multiple abscess formation with subsequent necrosis of the overlying skin resulting in multiple ulcers.

(b) *Syphilitic or Gummatous ulcer*: Occurs in tertiary stage of syphilis, as a result of necrosis of a chronic granuloma of the muscle.

(c) *Bairnsdale ulcer*: Starts as a chronic inflammation of the skin caused by the acid-fast bacilli known as *Mycobacterium ulcerans*. First recognised in Australia but common in Uganda where it is known as *Buruli ulcer*.

Neuropathic ulcer

The basic defect is sensory loss due to neurological disorder. The ulcer occurs due to repeated injury or pressure which is allowed to occur *because of loss*

of appreciation of pain in the area. These ulcers are also called *trophic ulcers or penetrating ulcers*.

The ulcer commonly occurs on the sole of the foot or on the heel and it may penetrate the bone or joint.

Common conditions which predispose to neuropathic ulcers are diabetes or alcoholic neuritis, tabes dorsalis, syringomyelia and paraplegia.

In diabetes mellitus, the ulcer may be precipitated also by atherosclerosis and infection. The toes and the feet are commonly affected.

Neoplastic ulcer

Primary tumour of the skin, e.g., squamous cell carcinoma, malignant melanoma may present as an ulcer.

Metastasis from a distant primary focus can be lodged in the skin and may undergo necrosis and ulceration.

Marjolin's ulcers: Malignant change may occur in long-standing venous ulcers, or in the scars of old ulcerated burns which refuse to heal, or in keloid, or in chronically discharging osteomyelitic sinuses.

Cryopathic ulcer

The term is used to describe *an ulcer which results from exposure to extremely low temperature.*

(a) *Chilblains (perniosis)*: Intense vasoconstriction of the skin arterioles in areas exposed to cold of moderate intensity results in tender, red, pruritic swelling followed by blister formation and subsequent ulceration. The lesions are focal and superficial which may heal rapidly. The lesions commonly affect the feet and toes of individuals particularly susceptible to cold injury.

(b) *Frostbite*: Exposure of a part of the body in the *wet cold* at above freezing temperature for a prolonged period results in ischaemic changes of skin and subcutaneous tissue due to intense arteriolar spasm. There is also stasis of blood in the damaged capillaries, which aggravates the ischaemia. Severe freezing injury may cause freezing of tissues, denaturation of intracellular protein and destruction of enzyme systems and promote ice-crystal formation in the cells. All these changes cause gangrene of at least the full thickness of the skin and subsequent ulceration.

Idiopathic ulcer

(a) *Tropical ulcer*: Commonly occurs on the legs and feet of the people living in the tropical countries. Malnutrition is supposed to be a factor.

(b) *Martorell's ulcer*: Also known as *Hypertensive ulcer*, because it occurs in hypertensive patients. But usually there is no associated atherosclerotic peripheral arterial disease. The ulcer occurs in the leg, and to start with, there are patches of spontaneous skin necrosis.

Self-inflicted ulcer

Due to injury to the skin by scratching, cutting or the injection of substances; found most often in those who are psychologically abnormal or who hope for some personal gain.

DIFFERENTIAL DIAGNOSIS OF ULCERS OF THE LEG

A. Venous ulcer

1. Duration – long.
2. *Site*: It occurs typically over the medial aspects of the lower third of the leg around and above the medial malleoli because of the presence of large number of perforators in this area.
3. Ulcer is ovoid in shape with irregular margin. Usually single in number. The ulcer may be fixed to the bone due to associated periostitis. The ulcer may be at the healing stage when it has a slopping edge and a base of healthy granulation tissue. The ulcer may be painful and tender.
4. *Presence of associated varicose veins*.
5. *Presence of unhealthy, pigmented skin* (bluish in colour) with or without oedema of the leg.
6. Presence of deformity (Equinus) and disfigurement (bottle-neck appearance).
7. (a) Evidence of perforator incompetence, Pratt's Test – Positive.
(b) Homan's sign (Passive dorsiflexion of the foot will cause intense pain in the calf) – if positive, suggestive of deep vein thrombosis.
(c) Perthes's test – may be positive.
8. *Peripheral arterial pulses are normal* (cf. Arterial ulcer).
9. Investigations – Venography may help, if deep vein thrombosis and perforator incompetence are suspected.



Fig. 17.1. Varicose ulcer on the medial side of lower 1/3rd of the leg. Note the pigmentation, the ankle flare, and varicosity at the upper half of the leg. All these findings led to the diagnosis.

B. Arterial ulcer (syn. Ischaemic ulcer)

1. *Site*: Occurs on the toes, dorsum of the foot, or heel.
2. *History of intermittent claudication, rest pain* particularly in a person who is a chronic smoker.
3. Arterial ulcers are very painful.

On examination:

4. The arterial ulcer varies in size and shape. The edge is punched out because healing by the surrounding tissues is poor or absent as they too are ischaemic. The skin at the edge may be greyish-blue in colour.
The base may contain grey-yellow sloughing tissue and is often infected. The ulcer is very often deep penetrating down to the bone or into the underlying joints. Occasionally the bare bone, ligaments or tendons may be exposed at the base of the ischaemic ulcer. *The ulcer and the surrounding tissues are very tender.*
5. *Peripheral arterial pulses – Poor or absent.*



Fig. 17.2. Varicose ulcers. The upper one was healed. The lower one was in healing stage. Note the pigmentation around. No varicosities were noted nearby even after tourniquet test as this patient had Trendelenburg's operation with stripping of the great saphenous vein few years back. So, ask for the previous operation and look for the scars—at the groin and above the medial malleolus.

6. Presence of ischaemic changes in the limb like dry pale skin, loss of hair, fissuring of nails, etc. The surrounding tissues are usually cold because they too are ischaemic.
7. Arteriography – may help, if peripheral arterial disease is suspected.

C. Neuropathic ulcer

1. Site – occurs on the sole or heel of the foot.
2. Neuropathic ulcer is a deep penetrating ulcer like arterial ulcer but it is *painless and non-tender*. It occurs over the pressure areas. The surrounding tissues are healthy and have a normal circulation but are unable to appreciate pain.
3. Evidence of neurological deficits is present, e.g., loss or diminution of sensation, weakness of the muscles.
4. Peripheral pulses are usually present because diabetes affects the smaller vessels like digital vessels mainly.

5. Blood and urine for sugar – to see diabetes mellitus.
6. Blood for W.R. and Kahn – to exclude tabes dorsalis.

D. Malignant ulcer

See squamous cell carcinoma and malignant melanoma (Chapter 1).

1. The ulcer has a rolled out and everted margin.
2. Biopsy – must be done, if malignancy is suspected.

E. Traumatic ulcer

1. Site – commonly over the area where the skin is closely applied to bony prominences, e.g., shin, malleoli, the back of the heel, the back of the sacrum, etc.
2. The ulcer may be due to bed-sores or plaster sores.
3. The ulcer appears as a painful, tender small circular lesion.

F. Cryopathic ulcer

1. Site – commonly the toes are affected.
2. History of exposure to freezing or near freezing temperatures are present.
3. To start with there is blister formation followed by ulceration.

G. Gummatous ulcer (Syphilitic ulcer)

1. Duration – short.
2. Site: It occurs classically on the outer side of the leg and more commonly on the upper aspect, i.e., commonly round about the knee.
3. History of exposure.
4. The ulcer is typically of punched-out appearance with clear-cut vertical edges and wash-leather slough on the floor. The ulcer may be round or serpiginous in shape.
5. The ulcer is *painless*, and may be multiple in number.
6. Absence of varicosity.
7. Associated features of neurosyphilis like A.R.P. and cardiovascular syphilis like aortic incompetence, aneurysm of the arch of aorta may be present.
8. Blood for W.R. and Kahn – highly positive.

9. Culture of the ulcer discharge – may show treponema.

H. Bairnsdale ulcer

1. Site – can occur anywhere, commonly in the limbs.
2. *The ulcer is irregular with thin and undermined edges* with frequent presence of bluish, pale, watery granulation tissue on the floor.
3. Smear examination and culture of discharge – may show acid-fast bacilli.
4. Absence of varicosity.



Fig. 17.3. Tuberculous ulcer over the lateral malleolus of the ankle. Note the undermined edge of the ulcer.

MANAGEMENT OF LEG ULCERS

1. Careful history taking with particular emphasis on the following:
 - (a) Duration
 - (b) Painful or painless
 - (c) History of claudication
 - (d) History of diabetes
 - (e) History of D.V.T.
 - (f) History of exposure.
2. Local examination of the ulcer regarding the character of the ulcer – Site, shape, size, edge,

floor, base, any fixity to the bone.
Note for tenderness and induration of the ulcer margin or base.

Note for surrounding skin changes and for presence of varicosities.

3. Palpation of the peripheral arterial pulses.
4. Examination for any evidence of neurological disorders – Sensory or motor impairments.
5. Examination of the draining lymph nodes.
6. Investigations:
 - (a) Blood for routine, sugar and W.R. and Kahn.
 - (b) Urine for sugar.
 - (c) Culture of ulcer discharge for:
 - (i) Pyogenic organisms.
 - (ii) Mycobacteria.
 - (iii) Treponema.
 - (d) Arteriography – if peripheral arterial disease is suspected.
 - (e) Venography – if deep vein thrombosis and perforator incompetency are suspected.
 - (f) Biopsy – whenever a reasonable doubt exists as to the nature of the ulcer, a biopsy should be done.
 - (g) Therapeutic test – when self-inflicted ulcer is suspected, plaster immobilisation of the affected part will help in the healing of the ulcer, but amazingly it is found that the patient may deliberately create ulcer on some other parts of the body.
7. Treatment of the ulcer – varies according to the nature of the ulcer.

Causes of Delayed Healing of Ulcer

1. Old age.
2. Malnutrition: Hypoproteinaemia, avitaminosis, anaemia.
3. Secondary infection.
4. Diabetes mellitus.
5. Ulcer situated over the bony prominences.
6. Neurological deficit, e.g., sensory loss.
7. Excessive movement of the ulcer bearing area.
8. Callous ulcers (callous means hard or indurated).
9. Malignancy.

TREATMENT OF VARICOSE ULCER

- A. Conservative
- B. Operative

Conservative

Indications

1. Severe ulceration of long duration and the ulcer is associated with wide areas of skin involvement with dense fibrosis and brawny induration.
2. Presence of eczema which may be dry and coarse, scaly, moist or infected.
3. Pre-operative preparation before any definitive surgical treatment.
4. If operation is refused.

Principle

Principles are laid down according to *Bisguard regime* which advocates elevation and proper bandaging of the limb together with proper exercises and massage. By this regime almost any venous ulcer can be healed.

Methods

These depend on whether the ulcer is infected or not.

1. CLEAN ULCERS

- (a) Rest in bed with the foot end of the bed elevated by about nine inches.
- (b) Application of dressing and compression bandages.
 - (i) Application of zinc cream to the ulcer, olive oil to the dry surrounding skin, and calamine lotion to the eczematous areas that are itchy.
 - (ii) Dry gauze is then applied over the ulcer.
 - (iii) A felt pad or sponge rubber piece with bevelled edges, and cut to a size larger than the ulcer is then held over the ulcer by a crepe bandage.
 - (iv) Over this a firm one-way stretch elastic bandage is applied spirally from the base of the toes to the knee using a good one-half overlap with firm and even pressure, particularly around the foot and ankle. This type of bandaging helps to reduce oedema, to produce an added pumping and massaging effects of the leg muscle during walking and compression of the dilated veins and incompetent perforators.

Frequency of Application of bandages: The first application of bandage is removed after one week, and thereafter the bandage is renewed at fortnightly intervals until the ulcer is healed.

- (c) Massage: Frequent massage to the whole leg

in elevation may help to clear local oedema and to soften the indurated area round the ulcer.

- (d) Exercises – Both passive and active:

- Passive movements to maintain the mobility of the foot and ankle joint.
- Active movements of the calf muscles both in elevation and in standing (with bandages on).

(e) Walking training: The patient should be taught proper walking. The heel touches the ground first and then the calf muscles should be used to lift the heel of the back foot. This will give a 'spring' to the walk and thereby improve the venous pump.

Once the ulcer is healed by above procedures, the patient is instructed to wear an efficiently fitted rubberized cotton or nylon stocking, or a crepe bandage, from the base of the toes to the knee. The stocking or crepe bandage should be applied before rising from the bed every morning. The bandage may be discarded at night, or during the day for short periods, provided the foot end of the bed is elevated.

The limb should be kept elevated for a while after each walking. Calf muscle massage should be continued particularly in elevation.

2. INFECTED ULCERS

The idea of treatment is to convert an infected ulcer into a clean one which is then treated as above.

- (a) Swab for culture and sensitivity to antibiotics.
- (b) Application of Eusol and/or hydrogen peroxide to the ulcer to remove the slough and pus followed by dressing until the ulcer becomes dry with healthy pink granulations tissue at the base.
- (c) Antibiotics: Local application of antibiotics is unnecessary and may be harmful if a sensitivity reaction or a resistant organism develops.

Systemic antibiotics may be required in presence of cellulitis, lymphangitis and lymphadenitis.

- (d) *Rest the patient in bed with the leg elevated when the infective-process shows signs of spreading.*

Once the infection is controlled, the ulcer should be treated along the lines indicated for a clean ulcer.

Operative

Indications

1. Small ulcers with or without localised changes in the surrounding tissues.
2. Extensive ulcer, over 1 inch in diameter, with an induration of the surrounding area over 2

inches in diameter – when the healing is delayed or difficult to achieve or difficult to maintain by conservative methods.

3. When the ulcer is healed – to prevent recurrence.

Contraindications

1. Severe ulceration with wide areas of skin deterioration and/or diffuse brawny oedema, because there is a possibility of skin necrosis in these cases after a perforator dissection.
2. Presence of infected ulcers.
3. Presence of moist or infected eczema.

Methods

1. Subfascial perforator dissection and ligation (*Cockett and Dodd operation*).
2. Excision of associated varicose veins which are the cause of the ulceration or are contributory to ulceration.
3. Excision of the ulcer and also of the nearby unhealthy skin down to healthy fascia or muscle followed by skin grafting.

In order to avoid oedema, pressure bandages must be worn for a few weeks after operation.



18

CHAPTER

Swellings of the Leg

Swelling of the lower limb are often encountered in clinical practice. Commonly, they are due to medical conditions such as heart failure, kidney disease or malnutrition. But the surgeons are concerned only with the peripheral causes, the majority of which are venous or lymphatic in origin.

CLASSIFICATION OF CAUSES

Central

Swelling is usually bilateral.

1. *Cardiac*
 - Congestive heart failure
 - Constrictive pericarditis
2. *Renal*
 - Nephrotic syndrome
 - Acute nephritis
3. *Hepatic disorder* – multiple factors are responsible for oedema.
4. *Gastrointestinal*
 - Gastroenteropathies.
5. *Nutritional*
 - Protein deficiency
 - Thiamine deficiency
 - Iron deficiency
6. *Hormonal*
 - Cushing's syndrome
 - Myxoedema
 - Pre-tibial myxoedema in thyrotoxicosis

Peripheral

Swelling is usually unilateral.

1. *Venous disorder*
 - Perforator vein incompetency
 - Calf vein thrombosis
 - Iliofemoral vein thrombosis
 - Inferior vena cava thrombosis
2. *Lymphoedema*
 - A. Primary – due to developmental anomaly:
 - Lymphoedema congenita
 - Lymphoedema praecox
 - Lymphoedema tarda
 - B. Secondary – commonest cause of lymphoedema and is due to obstruction of previously normal lymphatic channels by a known agent such as:
 - (a) Iatrogenic:
 - Following extensive surgical removal of lymph nodes
 - Post-irradiation fibrosis.
 - (b) Malignant infiltration of lymph nodes and channels.
 - (c) Inflammation:
 - Lymphogranuloma inguinale
 - Tuberculosis
 - Recurrent non-specific infection.
 - (d) Parasitic infestation of lymph nodes, e.g., filariasis.

3. Miscellaneous

- Adiposis dolorosa
- Erythrocyanosis frigida
- Arteriovenous fistula
- Tight plaster or bandage
- Injuries – fracture, muscle contusion, extensive burn
- Infection – cellulitis, abscess, gas gangrene.

SURGICAL PHYSIOLOGY

Normal tissue fluid circulation

Normally, there is a continuous circulation of fluid out of the arterial end of the capillaries into the interstitial spaces and then back into the venous end of the capillaries. In order to maintain this normal circulation of fluid across the capillary wall a balance between two main forces, i.e., hydrostatic pressure or capillary blood pressure and plasma osmotic pressure, operates which controls the rate and direction of fluid movement (Fig. 18.1).

Some fluid and most proteins ultrafiltrated at the level of the arterial capillaries enters the lymphatics before eventually returning to the circulation. This may be partly the result of tissue pressure and partly due to osmotic attraction of proteins in the lymphatic system.

Oedema is defined as the swelling of the tissues, caused by an increased interstitial fluid. *Venous obstruction reduces* the quantity of fluid reabsorbed at the venous end of the capillaries resulting in oedema containing a large quantity of watery interstitial fluid. *Lymphatic obstruction reduces* the

clearance of interstitial protein along with water, and produces the oedema containing interstitial fluid with a high protein concentration. *The main difference between venous and lymphatic oedema is the protein concentration of the interstitial fluid and the rate of increased interstitial fluid formation in the latter.*

So estimation of protein content of oedema fluid gives a useful guide to the nature of the oedema. *In venous oedema* (and also in cardiac and hypoproteinimic oedema), the protein content is usually under 1.0 gm percent, *while in lymphoedema* (and also in situations of increased capillary permeability following allergy and burns), the protein content is 1.5 gm percent or over.

SURGICAL PATHOLOGY

Oedema from venous disorder

Following engorgement of the superficial venous system of the lower limb due to perforator vein incompetence or thrombosis of the deep venous system, the hydrostatic capillary pressure of blood exceeds its plasma osmotic pressure. As a result, the fluid of low protein content accumulates in the interstitial spaces causing *pitting oedema of the limb*.

Extensive oedema of the whole leg is seen after massive thrombosis of whole deep venous system of the leg, the iliofemoral veins (phlegmasia caerulea dolens), or the inferior vena cava. Thrombosis of the inferior vena cava may occur either as an extension from the iliofemoral thrombosis, or in association with abdominal cancer, puerperal sepsis or other infective process. *The oedema then is usually bilateral.*

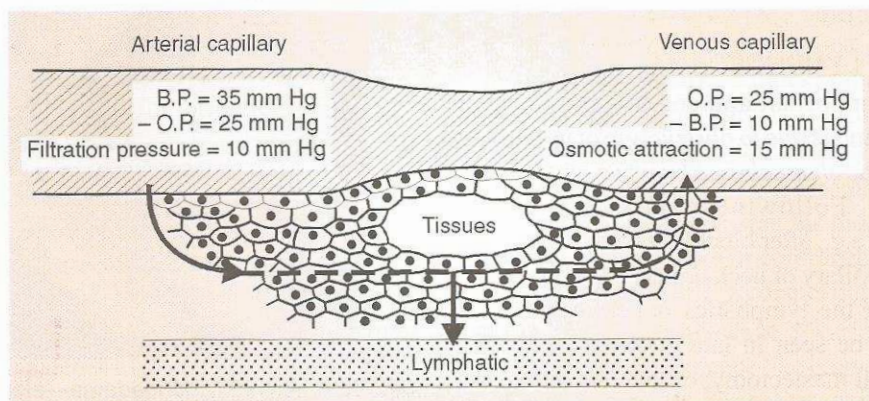


Fig. 18.1. Normal rate and direction of fluid movement.

Lymphoedema

Lymphoedema is interstitial oedema of lymphatic origin resulting in protein-rich fluid accumulation in the interstitial compartment. It results from the obstruction of lymphatic flow due to fault in the lymphatic system. This fault may be primary or secondary.

PRIMARY LYMPHOEDEMA

It occurs due to developmental defects of the subcutaneous lymphatic channels.

Lymphangiography demonstrates three types of defect in the lymphatic system.

1. *Aplasia*: Where the lymph channels are not formed at all.
2. *Hypoplasia*: Where the lymph channels are too small or too few.
3. *Varicose*: Where the lymph channels are dilated, tortuous and incompetent. When these varicosities extend into the pelvic and para-aortic lymph trunks, retrograde flow of intestinal chyle may occur into the groin and thigh causing chyle-filled vesicles to appear beneath the skin.

Primary lymphoedema is described in 3 clinical sub-groups according to the age of onset:

Lymphoedema congenita (10%): The swelling is either present at birth or appears shortly afterwards. When familial, the condition is known as *Milroy's disease*.

Lymphoedema praecox (75%): The swelling usually appears in childhood or adolescence. It accounts for by far the largest groups.

Lymphoedema tarda (15%): The swelling appears in adult life.

SECONDARY LYMPHOEDEMA

It is the most important and commonest cause of lymphoedema and is *due to obstruction of previously normal lymphatic channels by a known agent such as*:

1. Trauma – Following extensive surgical procedures e.g., after block dissection of the ilio-inguinal, axillary or neck nodes where extensive removal of the lymphatics is performed. This may often be seen in late oedema of the arm after radical mastectomy, or late oedema of the limbs after radical surgery in malignant melanoma.
2. Malignancy – Advanced neoplastic infiltration of the lymph nodes causes lymphatic blockage, e.g., brawny arm or cancer-en-cuirasse in breast carcinoma.
3. Parasitic infestation e.g., Filariasis. The *Wuchereria bancrofti* infects the lymphatics; a chronic and recurrent inflammatory reaction is set up with consequent lymphatic obstruction leading to gross lymphoedema. It usually occurs in the lower limbs and genitalia.
4. Post-irradiation fibrosis.
5. Inflammation e.g., repeated attacks of streptococcal cellulitis may cause fibrosis of lymphatic vessels, common in those who go about barefoot.

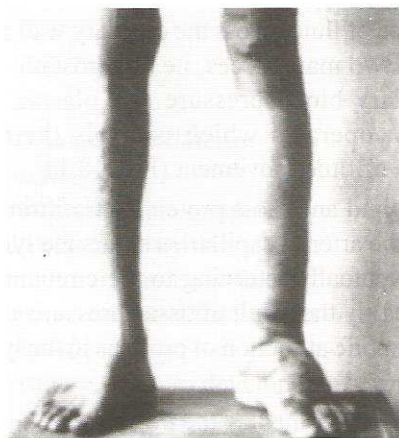


Fig. 18.2. Unilateral left leg swelling due to filariasis.

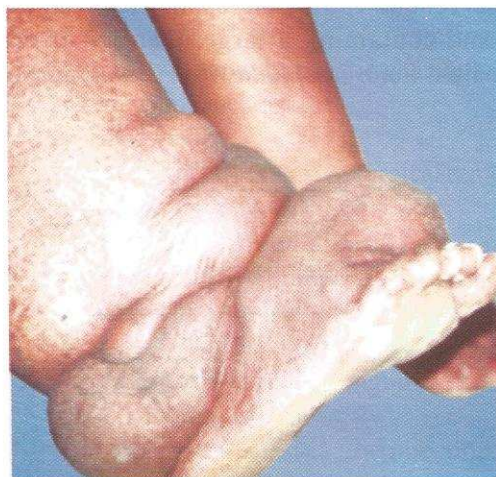


Fig. 18.3. Filarial lymphoedema—elephantiasis of the foot and leg. Grotesque appearance. (Courtesy: Dr. Yogesh Salphale, D'ortho)

Sequence of events in lymphoedema

Whatever may be the cause, following obstruction, the lymphatic vessels fail to remove the protein molecules and larger particles adequately from the tissues. As a result, there is an increase in tissue colloidal osmotic pressure which increases filtration of fluid across the capillary membrane and decreases reabsorption so that fluid of higher protein content accumulates in the interstitial space leading to soft pitting oedema. *In the second stage*, as the process continues and becomes chronic, the skin and subcutaneous tissues become thickened from fibrous tissue replacement and thus pitting is lost. *In the third or more advanced stage*, as the chronicity persists, there is excessive subcutaneous fibrosis and scarring leading to induration. The swollen limb becomes firm to hard in consistency with thickened skin. The dorsum of the foot is characteristically swollen like buffalo hump (Fig. 18.3) and the toes become thick and squared. Later still, the skin becomes hyperkeratotic and horny. Vesicular eruptions appear which ulcerate and discharge clear or milky (chylous) lymph. These changes are accentuated earlier when recurrent cellulitis occurs.

Miscellaneous causes

Adiposis dolorosa

There is abnormal and painful accumulation of fat particularly around the thigh, buttocks and ankles.

Arteriovenous fistula (see Chapter 15)

Congenital variety may be localised or diffuse. The diffuse variety is often associated with a giant swollen limb, varicose veins and varicose lymphatics, superficial angiomas in the adolescent.

Traumatic fistula occurs following penetrating injuries. The limb gradually swells and becomes associated with varicose vein formation.

Erythrocyanosis frigida

This condition is due to hypoplastic microcirculation of the skin arterioles, but the main blood vessels remain patent. The condition is almost entirely confined to female adolescents. Following exposure to cold, the skin of the ankles and legs becomes discoloured with patches of deep cyanosis alternating with reddened areas. The legs become permanently swollen with thickened subcutaneous tissue, and

subcutaneous fat necrosis may result in the formation of hard and tender nodules. Occasionally the nodules break down, leaving superficial ulcers which are very painful. *But the peripheral pulses are palpable.*

DIAGNOSIS

The cause of swelling of the leg can be established by careful history and physical examination. The surgeon is more concerned with the peripheral causes.

Is the swelling central in origin?

Oedema of central origin is usually generalised and the leg swellings are bilateral. Central causes must be excluded by careful history and physical examination.

Is the swelling unilateral in distribution?

Unilateral swelling of the leg usually indicates a local cause, which is of venous or lymphatic origin in the majority of cases; venous pathology is most common.

Is there any history of pain during development of swelling?

Oedema of central origin or lymphoedema is usually painless. Oedema of venous origin is usually painful.

Is the swelling venous or lymphatic in origin?

1. Venous origin

(a) History of deep vein thrombosis with the development of a painful swollen leg following operation, prolonged confinement to bed or pregnancy.

(b) History of massive deep vein thrombosis (*phlegmasia caerulea dolens*) with the development of acute, severe pain over most of the limb in association with marked shock, rapidly developing extensive oedema, cyanosis, turgid subcutaneous veins and superficial skin ulceration.

(c) Occasionally, deep vein thrombosis may occur without any apparent reason; the hidden malignancy or the prolonged intake of contraceptive pill may be considered as possible predisposing factors.

(d) There may be presence of varicose veins, signs of perforator incompetence like blowouts, fascial pits, ankle flare, varicose dermatitis, skin pigmentation, varicose ulcer.

(e) There may be presence of pitting oedema in the early stage. The oedema is greatest at the legs and ankle *with sparing of the foot*. The oedema reduces promptly to overnight elevation of the limbs. The oedema may become non-pitting when the swelling becomes chronic and associated with skin and subcutaneous tissue thickening.

II. Lymphatic origin

The patient presents with a history of slowly progressive swelling of the limb with or without the affection of genitalia. *The swelling is rarely painful at any stage*. The skin of a lymphoedematous limb remains remarkably healthy for many years apart from recurrent attacks of cellulitis. Skin pigmentation, skin ulceration are usually absent. Oedema is pitting in the early stage but becomes non-pitting in the chronic stage when skin and subcutaneous tissue thickening is present. Eventually deformity i.e., elephantiasis is developed.

(a) **Primary lymphoedema:** There may be family history in 20 percent cases (*Milroy's disease*). The swelling is unilateral in 50 percent cases. Usually the swelling is of gradual onset and becomes worse in warm weather. In congenital lymphoedema the swelling is either present at birth or appears very shortly afterwards. Primary lymphoedema of all three types is much more common in females.

When the swelling persists for many years, hyperkeratotic, horny and vesicular changes may occur which may lead to ulceration and discharge of lymph. In the rare cases of lymphoedema, vesicles of milky fluid may appear due to chylous reflux.

(b) **Secondary lymphoedema:** The swelling is of rapid onset and unilateral in distribution. Most often there is an obvious cause for its occurrence, e.g., scar mark due to groin dissection, history of radiotherapy, or malignant infiltration.

Lymphoedema and oedema secondary to ilio-femoral thrombosis are very often secondary to malignant disease. So, a thorough examination of the lymph nodes in the groin and other areas, inspection of the skin of the lower half of the body, a full examination of the abdomen to see any lump or enlarged nodes, inspection and palpation of genitalia, e.g., testes and rectal and vaginal examination are essential.

The lymph nodes draining the oedematous area will not be enlarged in primary lymphoedema, but may be enlarged and hard if they are infiltrated with malignancy.

It must be stressed that one should examine the whole patient, particularly the heart, abdomen, veins of the limb and lymph nodes. One should exclude central causes of oedema e.g. the cardiac and renal causes of oedema before considering the local causes, venous disorder or lymphoedema.

Is the swelling arteriovenous in origin?

On rare occasions, when central and local causes of oedema are excluded, an arteriovenous fistula may have to be considered. (See chapter on Aneurysm).

INVESTIGATIONS

When the diagnosis is still dubious some special tests may be helpful.

1. **Venography:** The injection of radio-opaque dye into an ankle vein, femoral vein, or intraosseous vein at iliac crest will indicate the site and extent of a thrombotic plaque.

2. **Arteriography:** When arteriovenous fistula formation is suspected.

3. **Duplex ultrasound,** a modality that combines doppler ultrasound and colour flow imaging, is useful for diagnosis of DVT, and arteriovenous fistula.

4. **Lymphangiography:** Dorsal foot lymphatics are visualised following injection of a diffusible dye known as patent Blue V into the interdigital web spaces, 0.2 ml being injected into each space. Then a suitable lymphatic trunk at the dorsum of the foot is exposed under general anaesthesia and injected with radio-opaque solution such as Ethiodol and then multiple X-rays are taken over the next 24 hours to outline the nature of the lower limb and abdominal lymphatics and lymph nodes. Recently, isotope lymphangiography using radio-labelled (Technitium 99m) colloid is used for functional assessment of the lymphatic system.

5. Recently, high resolution ultrasound scan, CT scan or MRI have replaced lymphangiography to detect malignant lymph nodes.

TREATMENT

VENOUS DISORDER

Conservative

It should be followed in every case.

1. *Elevation*: Elevation of the limb helps to reduce the swelling. In the beginning it should be prolonged and continuous and when the patient is allowed out of bed he should arrange the intermittent elevation of the leg once during the day, and at bed time. He should avoid prolonged sitting or standing and should rest his legs upon a chair whenever he sits. While in elevation the patient should perform active movements and exercises of the limb.
2. *Elastic stockings*: Patients should wear well-fitted elastic stockings at all times, except when in bed. It helps in reducing the oedema, and counteracts the high venous pressure which develops during exercise. In absence of elastic stockings, crepe bandage may be applied firmly.
3. *Massage*: Massage in a centripetal direction, gently and firmly with lanolin or olive oil helps to push the fluid out of the subcutaneous tissue and keeps the skin and subcutaneous tissue soft.
4. *Anticoagulant therapy*.
5. *Fibrinolytic therapy* – if anticoagulants are contraindicated.
6. *Diuretics* may be used intermittently.

Surgery

When the conservative regimens fail to improve the oedema:

1. If seen in the early stage (i.e., within 48 hours of onset) – in addition to above measures some advocate thrombectomy in deep vein thrombosis.
2. If the obstruction is very short – the obstructed portion can sometimes be resected followed by end-to-end anastomosis.
3. *Palma operation*: Localised obstructions in the iliac vein can be bypassed in which the long saphenous vein of the healthy leg is rerouted subcutaneously across the pubis and anastomosed to a vein below the block in iliac vein.
4. Ligation and removal of superficial venous system e.g., long saphenous vein can be performed if the deep veins are healthy.

5. Perforator vein incompetence, venous ulceration may be improved by subfascial perforator dissection and ligation (Cockett and Dodd operation).

LYMPHOEDEMA

Conservative

It should be followed in every case.

- | | |
|----------------------|------------------------------------|
| 1. Elevation | } Discussed under
venous oedema |
| 2. Elastic stockings | |
| 3. Massage | |

For lymphoedema, a high quality elastic compression garment is worn by the patients, preferably at all times except when the limbs are elevated above the heart. Such garment also protects the limbs from injuries such as burns, lacerations and insect bites and infections.

4. Great care in the hygiene of the affected part:
 - (a) regular cleaning preferably with hexachlorophene soap or savlon solution;
 - (b) many of these patients have Tinea pedis, so, the regular use of fungicidal ointment helps to reduce the incidence of cellulitis. In bad cases, oral griseofulvin may be added;
 - (c) use of antibiotics, if bacterial infection persists – mostly the organisms are streptococcus, so, penicillin, or erythromycin in penicillin sensitive or resistant cases, is preferred.
5. Diuretics may be used intermittently.
6. Intermittent pneumatic compression throughout the night can often help to reduce the oedema, but it causes sleep disturbance.
7. Benzopyrones (e.g. Coumaran) are being used in USA and Europe. They are thought to reduce lymphoedema through stimulation of proteolysis by tissue macrophages and stimulation of peristalsis and pumping action of the collecting lymphatics. Benzopyrones have no anticoagulant action.

Surgery

Indication

1. Gross swelling not responding to conservative measures. Gross swelling incapacitate the individual due to the weight and bulk of the limb causing limitation of movements.

2. Cosmetic reasons.
3. Chronic, neglected cases with recurrent cellulitis.

Types

- Direct
- Indirect.

Direct surgery

Direct attack to the lymphatic system is rarely performed. Only in a few cases where lymphangiography shows reflux of chyle downwards from the abdominal lymphatics to those of the leg – the reflux can be reduced by ligature and division of the dilated lymphatics which are usually approached in the inguinal region.

Indirect surgery

- (a) Excisional or reducing operations.
- (b) Reconstructive operations to provide new lymphatic pathways.
- (c) Amputation.

Reducing operations

There are three types of reducing operations available.

1. *Partial subcutaneous tissue excision* (Homans): In this operation a long skin flap is reflected along the axis of the limb and the subcutaneous tissues are excised from one half of the limb. The skin is closed with simple suture. The procedure is repeated on the other half of the limb three months later.

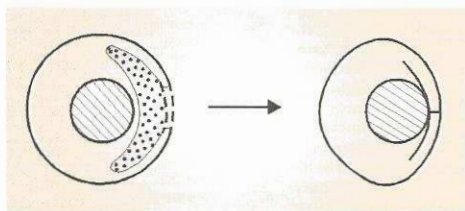


Fig. 18.4. Partial subcutaneous tissue excision (Homans).

2. *Excision plus buried dermis flap* (Thompson): Also known as Swiss roll operation. This is an extension of the partial excisional procedure in which one of the skin flaps is shaved of its superficial layers

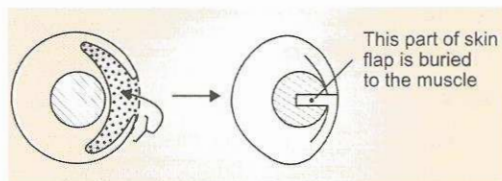


Fig. 18.5. Excision plus buried dermis flap (Thompson).

of epidermis and then buried deep to the deep fascia. In this way new connections would develop between the dermal lymphatic plexus and the deep lymphatics.

3. *Total excision of grossly oedematous tissue* (Charles): This consists of a total excision of the skin, the whole of the oedematous subcutaneous tissue down to the deep fascia followed by resurfacing of the raw area with split skin grafts. The skin grafts could be taken either from the healthy part of the

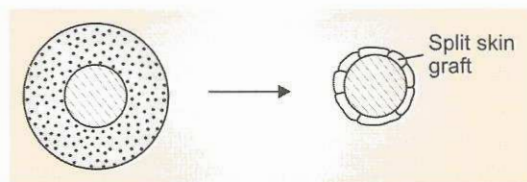


Fig. 18.6. Total excision and skin grafting (Charles).

body or from the excised skin. This is the most effective operation when there is gross oedema and the skin is healthy and non-infected.

Reconstructive operations

To provide new lymphatic pathways:

1. Lymphovenous shunts.
2. Transfer of lymphatic bearing tissue (portion of greater omentum) into the effected limb with hypoplastic and fibrotic distal lymphatic vessels in patients with primary lymphoedema.
3. Alternatively, a segment of ileum is disconnected from the rest of the bowel and stripped of its mucosa. Then this denuded ileum is mobilised and sutured onto the cut surface of the residual ilioinguinal nodes in an attempt to bridge the lower extremity with mesenteric lymph nodes.

Lymphovenous shunts

The lymphovenous shunts are mostly tried for secondary lymphoedema. The lymphatics can be joined to a vein in three ways:

1. By anastomosing a transected lymph node to a large vein (Fig. 18.7A).

2. By threading lymphatics into a vein (Fig. 18.7B).
3. By microsurgical anastomosis of the dilated lymphatics to a vein (Fig. 18.7C).

But none of these operations are satisfactory.

Amputations

May be required rarely for severe lymphoedema with skin ulceration and leakage of lymph, particularly when it incapacitates the patient or leads to a grotesque look of the limb.

ARTERIOVENOUS FISTULA

See Chapter 14.

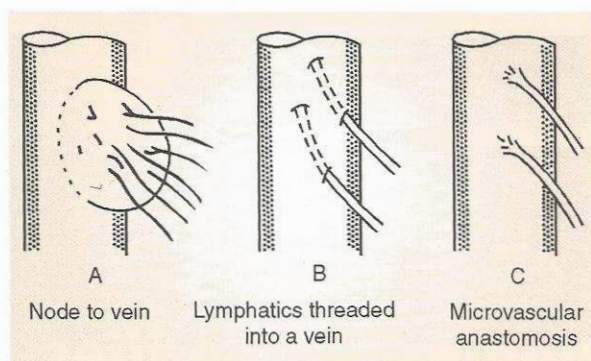


Fig. 18.7. Methods of lymphovenous shunts.

19

CHAPTER



Congenital Anomalies

CONGENITAL ANOMALIES OF THE VERTEBRAL COLUMN AND THE SPINAL CORD

Development of the spinal cord

Spinal cord develops from the neural tube.

1. In the early first week of intrauterine life, a dorsal groove appears on the surface ectoderm of the embryo – the neural groove.

2. The neural groove gradually deepens and ultimately becomes closed off forming the neural tube. From the walls of this neural tube the entire central nervous system (including spinal cord) is developed. Its lumen persists as the central canal.

3. The neural tube becomes separated from the surface ectoderm by the dorsal ingrowth of the surrounding mesoderm.

Development of the vertebral column

1. Anterior to the neural tube there is appearance of a solid rod of cells, the notochord, in the mesoderm. Round the notochord, the vertebral bodies develop.

2. From each side of the vertebral body two projections, one on each side, appear and they are known as neural arches.

3. The neural arches grow posteriorly to encircle the neural tube and ultimately fuse in the mid-line behind the neural tube to form the spine. The pedicles, laminae are developed from the neural arch. The transverse process is also developed from the neural arch as a lateral extension at the junction of pedicle and lamina.

4. *The fusion of the neural arches occurs first in the mid-thoracic region. From there fusion extends up and down.*

Developmental anomalies

Failure of the fusion of the neural arches gives rise to spina bifida and its variants.

1. Spina bifida (Fig. 19.1A).
2. Meningocele (Fig. 19.1B).
3. Meningomyelocele (Fig. 19.1C).
4. Syringomyelocele (Fig. 19.1D).
5. Myelocele (Fig. 19.1E).

The above mentioned defects can occur anywhere in the dorsal mid-line along the neuroaxis but are most common at its extremes, namely the lumbosacral and occipital regions.

SPINA BIFIDA (Fig. 19.1A)

There is a failure of fusion between the neural arches only. So the spinous process will be divided into two,

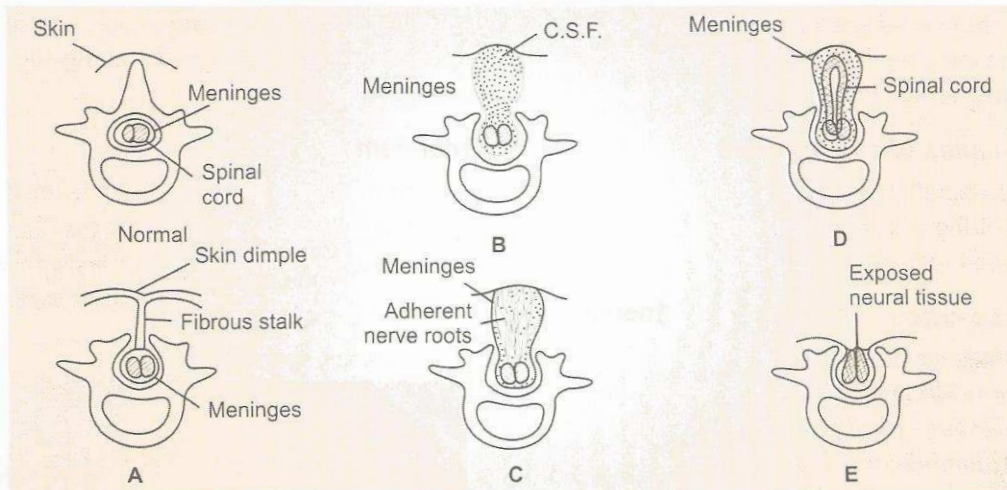


Fig. 19.1. Different anomalies of vertebral column and the spinal cord. A, Spina bifida occulta; B, Meningocele; C, Meningomyelocele; D, Syringomyelocele; E, Myelocele.

leaving a bony gap giving rise to spina bifida. Meninges and nervous tissue develop normally. The overlying skin usually remains intact, hence the deformity cannot be seen from the outside and then it is better called *spina bifida occulta*. The deformity is usually common in the lumbar or sacral region.

Diagnostic criteria

1. Present since birth.
2. A depression in the skin (skin dimple) – at the site of the defect.
3. Presence of a tuft of hairs over the defect.
4. Sometimes there may be presence of lipoma over the defect.
5. On palpation – a bony gap or bony indentation can be felt. Note the site of the defect in relation to bony landmarks.
6. Other congenital anomalies may be present e.g., cleft lip, phimosis, talipes, etc.
7. Neurological manifestations – if any, usually, do not appear before adolescence.
8. X-ray – will show the bony defect.

Explanation of the neurological manifestations

During birth the lower end of the spinal cord and its meninges are connected by a fibrous strand to the

skin of the back, known as *Membrana reuniens*. At birth the lower end of the spinal cord extends up to the coccyx. As the child grows the vertebral column grows more quickly than the spinal cord as a result of which the lower end of the spinal cord is apparently raised up and lies at the level of first lumbar vertebra. The membrana reuniens is thus made taut and stretched maximum when the spinal column has attained maximum growth in adulthood. This stretched membrana reuniens exerts traction over the meninges and the spinal cord to cause neurological manifestations, such as backache, nocturnal enuresis, paresis or even foot drop.

Many cases of spina bifida occulta are symptomless and so remains undiagnosed. They may be diagnosed by accident when an X-ray is taken for some other reasons.

Treatment

Minor degrees of spina bifida do not need any treatment, except for cosmetic reason to remove the tufts of hair or lipoma.

MENINGOCELE (Figs. 19.1B, 19.2, 19.3)

There is protrusion of the meninges through the gap in the neural arch posteriorly giving rise to a cystic swelling containing cerebrospinal fluid.

Usually the pia and arachnoid protrude but the dura stops at the margin of the defect. The overlying skin remains intact (Fig. 19.1B).

Common sites of meningocele

1. Lumbosacral region.
2. Root of the nose (cf. dermoid).
3. Occipital region (cf. dermoid).

Diagnostic criteria

1. Present since birth.
2. Cystic swelling present at the typical site.
3. Fluctuation – positive.
4. Transillumination – highly positive.
5. Swelling is compressible.
6. Presence of impulse when the child cries or coughs.
7. Overlying skin is free and may be thin.
8. The edge of the bony defect or bony indentation at the margin of swelling is palpable (cf. Dermoid cyst). *Note the exact bony landmark.*
9. May be associated with other congenital anomalies, but *neurological manifestations are usually absent.*

Complications

1. Rupture
2. Infection
3. Ulceration
4. May be associated with hydrocephalus when the condition is known as *Arnold-Chiari syndrome*. If hydrocephalus is present it requires treatment first before excision of meningocele.

Investigation

1. X-ray – will show the bony defect.
2. CT scan of the brain to exclude hydrocephalus.
3. MRI when the swelling is at the lumbosacral region or the craniovertebral junction to note:
 - Clear delineation of the swelling.
 - Relation of the swelling to the neural structures.
 - Cord tethering.
 - Position of the lower end of conus medullaris.
 - Associated Arnold-Chiari malformation in craniovertebral lesion.

If associated with hydrocephalus, excision of meningocele or meningocele will aggravate the

hydrocephalus. So, a ventriculoperitoneal shunt is to be performed before undertaking the repair of meningocele.

Treatment

Excision of the sac and closure of the meninges.

The bony gap is overlapped by the apposition of adjacent erector spinae muscles and the overlying fascia with the help of lateral release incisions. The skin is then closed.



Fig. 19.2. Meningocele thoracolumbar region – cystic compressible swelling at the midline of the back. Transillumination positive.



Fig. 19.3. Meningocele over the thoracic vertebral region.

MENINGOMYELOCELE (Figs. 19.1C, 19.4)

There is protrusion of the spinal cord or nerve fibres of cauda equina along with the meninges.

Features

- (a) This is the most common variety.
- (b) It most commonly occurs in the lumbosacral region.



Fig. 19.4. Meningomyelocele over the lumbosacral region – note the parchment-like skin covering the neural placode.

- (c) The bony defect is felt at the margin of the swelling and usually extends over three or more segments. *Note the exact bony landmark.*
- (d) The nervous tissue may be adherent to the undersurface of the sac.
- (e) *Neurological deficits are almost always present in the form of both sensory and motor deficits.* The motor deficit commonly affects the bladder and bowel sphincters, and the muscles of the lower limb which are flaccid in nature with absent reflexes. Sensation of lower limb is also impaired.
- (f) It may be associated with hydrocephalus.
- (g) It may be associated with other congenital anomalies particularly of the central nervous system and urinary tract system.

Diagnostic criteria

1. Presence of all the features of meningocele.
2. *Transillumination test* – will show opaque bands amidst the red glow. These opaque bands are due to the presence of nerve fibres.
3. Neurological deficit usually present, e.g., bladder or bowel incontinence, flaccid paraparesis, club-foot, etc.
4. X-ray – will show the bony defect. It may show other deformities of the vertebrae like hemi-vertebrae, scoliosis, kyphosis, etc.
5. CT brain to exclude hydrocephalus.
6. MRI.

Complications

1. Rupture
2. Infection
3. Ulceration

4. Urinary bladder dysfunction, e.g., infection, hydronephrosis, incontinence.
5. Musculoskeletal abnormalities including scoliosis, kyphosis, dislocation of hip – due to muscular hypotonicity and imbalance.
6. May be associated with hydrocephalus.
7. May be associated with Arnold-Chiari malformation.

Treatment

The treatment of patients with meningomyelocele includes treatment of the local condition and treatment of the associated problems like hydrocephalus, musculoskeletal abnormalities, and the bladder and bowel dysfunction.

Time of operation

It is advisable that the operation should be done within the first few hours of life. The points in favour of doing early operation are:

1. The sac is smaller in size.
2. The skin is more pliable.
3. There are fewer adhesions between the nervous tissue and the sac.
4. The chance of infection is minimum as the skin flora have not yet been established.
5. The neurological deficit can be reduced to a minimum (as afterwards the spinal cord becomes more prone to infection or drying or infarction. The infarction is due to kinking of the blood supply to the spinal cord as the sac is liable to grow out of proportion to the growth of the child due to distension by the fluid).
6. As the sac enlarges, the overlying skin will become atrophic and ulcerate.

Principles and steps of operation

1. Operation is performed under local anaesthesia.
2. The child is tied on a splint with the face downwards and head-side is kept at a lower level to minimise the escape of cerebrospinal fluid. The child is given a feeding-bottle during operation.
3. The sac is opened and the redundant membrane is excised.
4. If the neural elements are adherent to the sac, they are either freed by meticulous dissection or separated with a strip of attached membrane. Then the neural elements are replaced, within the spinal canal.

5. The membranes are sutured over the cord.
6. Adjacent spinal muscles are approximated in the mid-line over the bony defect.
7. The wound may be reinforced with flaps of deep fascia from the erector spinae muscle.
8. The skin is closed without any tension. If necessary, rotation flaps may be fashioned to make an adequate skin cover.

Post-operative management

1. Regular skull measurements for evidence of development of hydrocephalus. (Some cases may subsequently develop hydrocephalus from the effect of associated congenital abnormalities at a higher level. Occasionally the hydrocephalus gets arrested spontaneously. Otherwise, cerebrospinal fluid shunting procedure is to be carried out e.g., Ventriculoatrial Shunt).
2. X-rays of the skull, entire spine and hips, urine analysis, urine culture, intravenous pyelogram are to be carried out routinely at regular intervals. As well as consultation of urologist, orthopaedic surgeon, neurosurgeon and a physiotherapist must be necessary for long term care.

SYRINGOMYELOCELE (Fig. 19.1D)

There is protrusion of the spinal cord and the central canal of the cord is dilated to form a cavity filled with fluid.

Some neurological deficits are always present.

Clinically, very difficult to distinguish from meningomyelocele except that *transillumination test* is negative in syringomyelocele.

MYELOCELE (Figs. 19.1E, 19.5)

There is failure of development of the dorsal wall of the neural tube resulting in exposure of the spinal cord and its central canal to the exterior on the surface of the body.

There is a constant dribbling of cerebrospinal fluid through the defect.

The defect may be complete throughout the length of the cord or it may be partial. The partial variety is called myelocele.

Fortunately, the baby with this defect is still born. If the baby is born alive, death ensues within a few days from infection of the cord and meninges.



Fig. 19.5. Myelocele on the lumbosacral region – note the absence of parchment-like skin over the neural placode.

N.B. In every case of spinal defect – Neurological examination should be done routinely to detect any sensory or motor deficits.

CONGENITAL ANOMALIES OF THE LIP AND THE PALATE

Development of the face, lip and palate

About the 6th week of intra-uterine life, five processes appear around the stomodeum or primitive mouth at the anterior end of the embryo. These are:

1. Frontonasal process – single.
2. Maxillary processes – two, one on each side.
3. Mandibular processes – two, one on each side.

The frontonasal process arises from the capsule of the forebrain vesicle and descends like a curtain.

The frontonasal process divides into two lateral nasal processes and one median nasal process by the two olfactory pits. The median nasal process becomes bluntly bifurcated, forming two globular processes. The two globular processes ultimately unite to form the central depressed part of the upper lip or philtrum.

The two maxillary processes, one on either side appear from the dorsal ends of the mandibular processes of the first arches, and grow ventromedially.

The upper part of the maxillary process unites with the lateral nasal process to form the cheek, and the junction of these two processes indicates the position of the nasolacrimal duct. If there is a failure of fusion of these two processes, there will be a facial cleft.

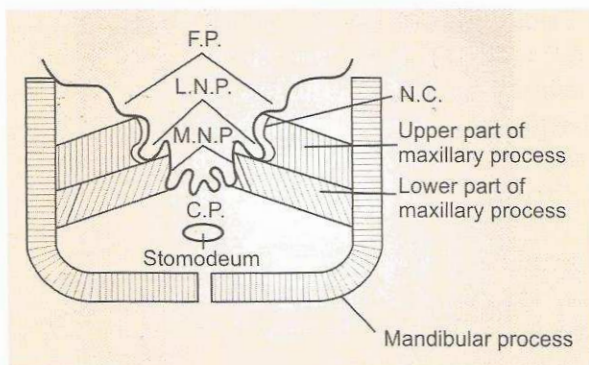


Fig. 19.6. Different processes taking part in the formation of face and hard palate. F.P., Frontonasal process; L.N.P., Lateral nasal process; M.N.P., Median nasal process; G.P., Globular process; N.C., Future site of nasolacrimal duct.

The lower part of the maxillary process grows medially beneath the olfactory pits, and unites with the median nasal process to complete the lateral parts of the upper lip.

So, the upper lip develops from two sources:

1. Philtrum – by the fusion of the two globular processes.
2. Lateral part of the upper lip – by the fusion of the median nasal process with the maxillary process.

Two palatine processes appear, one from each maxillary process. They grow downwards and medially beneath the olfactory pits (i.e., between the oral cavity and the nasal cavity) and ultimately fuse to form a part of the hard palate i.e., the primary palate.

On the anterior aspect of the palatine processes, there is a triangular gap which is filled up by a portion of the median nasal process known as pre-maxilla, with which the palatine processes fuse to complete the hard palate.

So, the hard palate develops from two sources:

1. From the primary palate which is developed by fusion of the two palatine processes – one on either side, derived from the maxillary processes.
2. From the secondary palate i.e., the pre-maxilla which is a part of the median nasal process.

The three segments of the palate unite around the incisive foramen. The fusion starts in front and proceeds backwards.

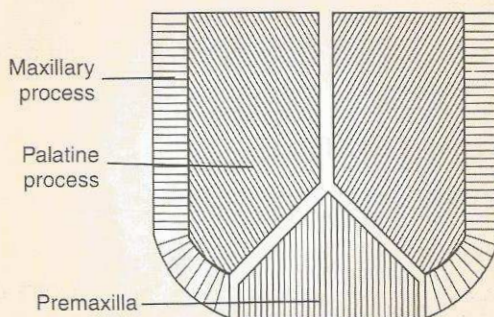


Fig. 19.7. Different parts taking part in the formation of hard palate.

From median nasal process and globular process develop:

- The philtrum of the upper lip.
- The pre-maxilla.
- The septum of the nose.

From maxillary process develop:

- The cheek.
- The whole of the upper lip except the philtrum.
- The palate.
- Most of the upper jaw.

From mandibular process develops:

- Lower jaw including the lower lip.

These processes unite around the stomodeum to form the face.

CONGENITAL ANOMALIES OF THE FACE

The anomalies can be classified in two groups—

I. Anomalies due to imperfect fusion:

1. Cleft lip:

(a) Upper lip:

- (i) Central
- (ii) Lateral

(b) Lower lip – Central.

2. Macrostoma and microstoma.
3. Cleft palate.
4. Facial cleft.

II. Anomalies due to inclusions of surface epithelium along the fusion lines:

1. Sequestration dermoids.
2. Fibro-chondromata.

CLEFT LIP

It is a gap in the lip, mostly congenital, but may be acquired due to trauma.

Congenital cleft lip

Commonly affects the upper lip.

Classification

1. *Central cleft*: It is exactly central and very rare. It is due to a failure of fusion of two globular processes derived from the median nasal process (Fig. 19.8A).

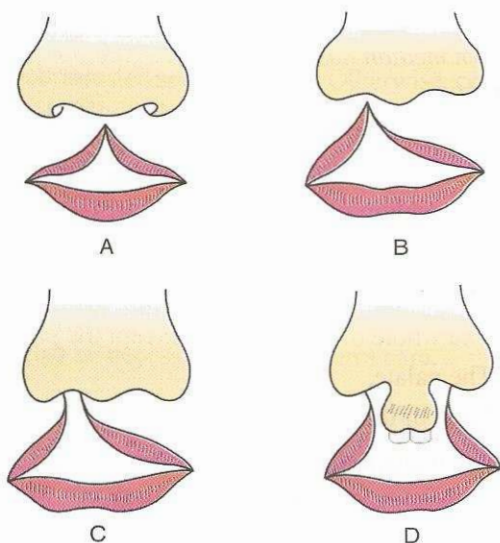


Fig. 19.8. Different types of cleft lip. A, Central cleft lip; B, Lateral cleft lip (incomplete); C, Lateral cleft lip (complete); D, Bilateral cleft lip.

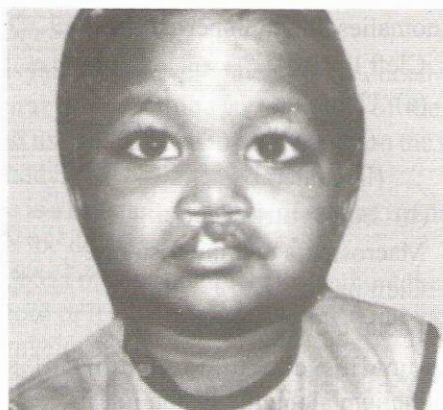


Fig. 19.9. Right lateral cleft lip—incomplete. The cleft not extending into the nostrils.



Fig. 19.10. Left lateral cleft lip—incomplete. The cleft not extending into the nostrils.



Fig. 19.11. Left incomplete cleft lip. Alveolus intact. But there is deformity of the left alae nasi.

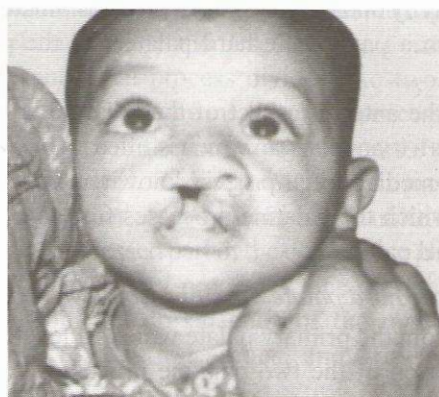


Fig. 19.12. Lateral cleft lip – complete and compound. The cleft extended into the nostril (complete). The cleft was associated with a cleft in the alveolus (compound). Look inside the mouth cavity to exclude cleft palate. Note the flattening of the right alae nasi. N.B. Look for congenital defects in other parts of the body.

2. **Lateral cleft:** Common type. The cleft is between the philtrum and the lateral part of the upper lip. It is due to failure of fusion between the median nasal process and the maxillary process. Lateral cleft lip again is classified as follows (Fig. 19.8).

(a) Unilateral (Fig. 19.8B & C) or bilateral (Fig. 19.8D).

Unilateral – cleft on one side of the philtrum. Bilateral – clefts on both sides of the philtrum. There may be forward protrusion of alveolus along with philtrum in bilateral complete cleft lip.

(b) Complete or incomplete (partial). It is regarded as complete when the cleft extends into the nostrils (Fig. 19.8C & D).

(c) Simple or compound. It is regarded as compound when the cleft lip is associated with a cleft in the alveolus (Fig. 19.12).

(d) Complicated or uncomplicated. It is regarded as complicated when the cleft lip is associated with a cleft in the palate (Fig. 19.14C).

Upper cleft lip may rarely be associated with the presence of two small blind tubes in the lower lip. These tubes are called inferior labial sinuses or mandibular recesses (Sutton). The tubes are lined with squamous epithelium. The cause is unknown, but there is a hereditary tendency.

CLEFT PALATE

It is a gap in the palate, mostly congenital.

Classification

Congenital cleft palate may be classified as follows:

Incomplete

1. Bifida uvula: Cleft of the uvula only (Fig. 19.13A).
2. Cleft of the soft palate along its entire length (Fig. 19.13B).
3. Cleft of the whole length of the soft palate and the posterior part of the hard palate. This is also known as intra-maxillary cleft palate and is due to a failure of fusion of the palatine processes (Fig. 19.13C).

Complete

Cleft on the soft palate and whole length of the hard palate, so that nose and mouth are one cavity.

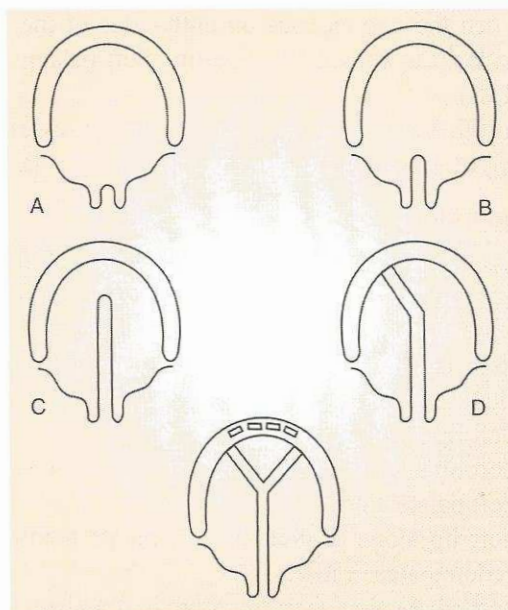


Fig. 19.13. Different types of cleft palate. A, Bifida uvula; B, Cleft soft palate; C, Intramaxillary cleft; D, Bipartite cleft; E, Tripartite cleft. A, B, C – incomplete; D, E – complete.

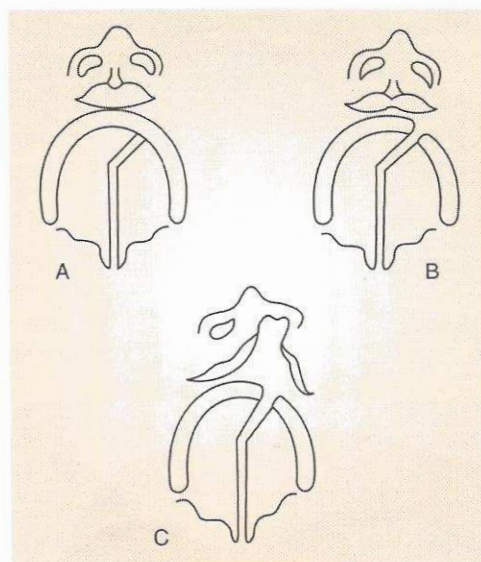


Fig. 19.14. A, Only cleft palate; B, Cleft palate and cleft alveolus; C, Cleft palate, cleft alveolus and cleft lip.

It is due to failure of fusion between the palatine processes and the pre-maxilla.

In front the gap may extend on one side of the pre-maxilla when it is known as Bipartite cleft palate (Fig. 19.13D, 19.14A).

When the gap extends on both sides of the premaxilla, it is known as Tripartite cleft palate (Fig. 19.13E).

In both these conditions the cleft may also extend to involve the alveolus and/or the lip (Fig. 19.14B, C).

Incidence

- The incidence varies in different countries and in different races. Incidence is *high in Oriental population and low in Negro population*.
- Cleft lip/palate occurs relatively frequently (1 in 800 live births).
- Cleft lip alone: 25 percent.
Cleft lip and cleft palate combined: 45 percent
Cleft palate alone: 40 percent.
- Cleft lip alone or cleft lip and palate combined predominates in males.
- Cleft palate alone appears more in females.
- Unilateral cleft lip is more common (75 percent).
- Left-sided cleft lip is the commonest variety.

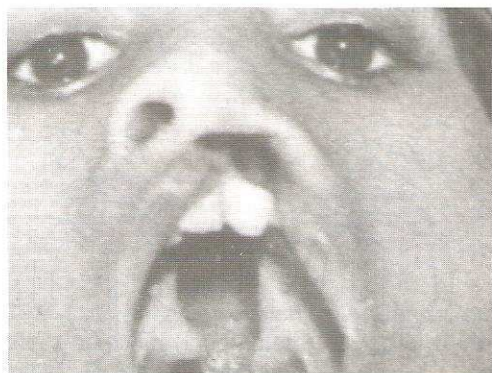


Fig. 19.15. Cleft lip + cleft alveolus (left side) + cleft palate. Note the flattening of alae nasi on the cleft side.

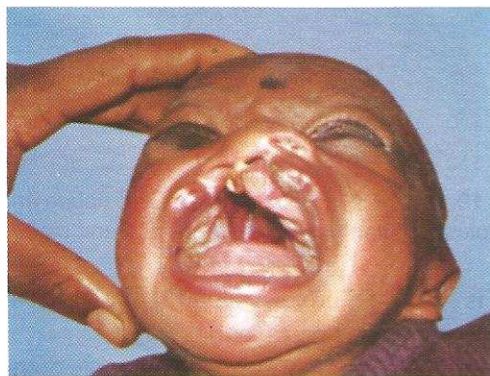


Fig. 19.16. Cleft lip + cleft alveolus (right side) + cleft palate. Note deformed alae nasi.

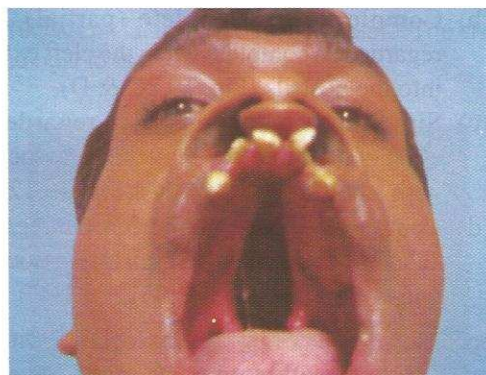
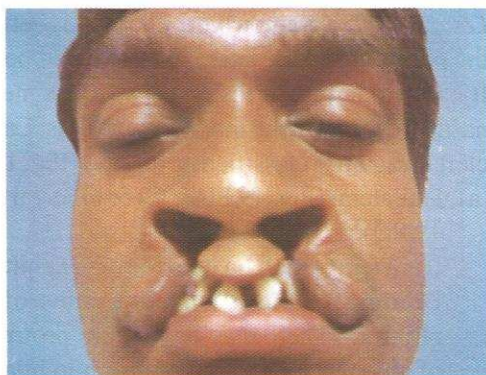


Fig. 19.17A&B Bilateral cleft lips + cleft alveoli + cleft palate. Note the symmetrical deformed alae nasi + protruding premaxilla.

- Incidence of bilateral cleft lip, incomplete or complete, is about 20 percent.

Aetiology of cleft lip / palate

- Both genetic and environmental factors are responsible.
- Genetic influence is more significant in combined cleft lips and palate than cleft palate alone where environmental factors exert a greater influence.
- Heredity may play a vital role. In 15 percent cases, the condition is familial. Hereditary transmission occurs through a male sex-linked recessive gene.
- Environmental factors are responsible during the first trimester of pregnancy when the mother suffers from one of the following:
 - (i) Nutrition deficiency, particularly Vit. B and Vit. A deficiency. Excess Vit. A may lead to congenital deformities. Antenatal folic acid supplement helps to prevent cleft lip/palate.

- (ii) Endocrine disorders.
- (iii) Certain viral infections, e.g., rubella.
- (iv) Excessive use of certain drugs, e.g., steroids, diazepam, phenytoin (in epilepsy).

Cortisone interferes with the synthesis of some essential amino acids and so with the protein metabolism in foetus. Cortisone also inhibits synthesis of Vit. B₆ and folic acid which are essential for nucleic acid production.

- (v) Anoxia – a threatened abortion may cause anoxia to the foetus and if the foetus is allowed to term it may be born with deformities.
- (vi) Exposure to radiation which is a potent teratogenic.
- (vii) Stress – excess of endogenous steroid secretion consequent upon stress.
- Maternal age: Children born at the late age of the mother are prone to suffer from congenital anomalies.
- Diabetes: Diabetic mothers are prone to give birth to deformed children.
- Consanguineous marriage: Also are believed to give birth to deformed children.
- Although most cleft lip/palate occur as an isolated deformity, *other syndromes may also be associated with it*, e.g., Down's, Apert's, Treacher Collin's, Stickler and Sprintzen syndromes, Pierre Robin's syndrome characterised by cleft palate, retrognathia and a posteriorly displaced tongue.

Diagnostic criteria

The patient is presented before the doctor by the parents mainly because of the deformity.

During examination of a case of cleft lip and/or cleft palate the following points should always be noted.

1. *What is the nature of deformity?* Cleft lip, cleft palate or both. So, during examination of a cleft lip, the palate should always be inspected (complicated or uncomplicated). Note the nature of the cleft in the palate (see classification).
2. *Site and side of the deformity* (see classification).
3. *Presence of any scar tissue* at the mucocutaneous junction of the lip – scar is absent in congenital cleft lip. Scar may be due to previous imperfect operation.

4. *Whether the cleft in the lip extends into the nostrils* (complete or incomplete).
5. *Whether there is any cleft in the alveolus* (compound or simple).
6. *Whether the alae nasi are flattened or not* – alae are usually found flattened on the side of the cleft.
7. *Position of the nasal septum.* Whether the nasal septum lies freely (complete cleft palate) or the nasal septum is attached to one side of the palate (incomplete cleft palate).
8. *Age of presentation:* If presented at a later age – the following points should be noted:
 - (i) the alignment of the teeth – whether regular or irregular?
 - (ii) forward protrusion of the alveolus;
 - (iii) any speech defect and nasal intonation;
 - (iv) any difficulty in swallowing or drinking;
 - (v) any evidence of chronic ear infection.
9. In addition, examination of the patient from foot to head has to be done for any other associated congenital deformities, e.g., undescended testis, hypospadias or epispadias, supernumerary digits, talipes equinovarus, defects in the spine, neurologic or cardiac anomalies.

Note: Cleft lip may be diagnosed in antenatal period by USG scan after 18 weeks of gestation; but cleft palate cannot be diagnosed antenatally.

Problems with cleft lip

1. Cosmetically it looks ugly.
2. Defective suction: Effective suction of breast is also helped by effective closure of the lips caused by the tight grip of the orbicularis oris muscle to create a vacuum (or negative pressure) inside the mouth. As there is a gap in the cleft lip, it results in an ineffective grip by the lip, which interferes with effective suction to some extent. So, nutrition of the newborn baby is, to some extent, hampered.
3. Defective dentition: Teeth tend to protrude through the gap along with the alveolus resulting in dental irregularity.
4. Defective speech particularly with the labial letters B, F, M, P, V, W (ওষ্ঠ্যবর্ণ – প্ ফ ব্ ভ ম – আভ্যবর্ণ প্ ফ্ ব্ ভ্ ম্)

Problems with cleft palate

1. Defective suction: For effective suction a vacuum (or negative pressure) is necessary inside the mouth, which cannot be established in cleft palate due to presence of an abnormal gap between mouth cavity and nasal cavity. Therefore, a baby suffers from malnutrition. So, in these cases feeding may be assisted by employing spoon feeding. In some cases an obturator may be applied to facilitate sucking.
2. Defective speech: Particularly with the palatal consonants (তালব্য বর্ণ / तालव्य वर्ण). There will be nasal intonation. To utter these letters there must be a proper closure of the nasopharyngeal isthmus also so that air must not escape into the nose via the nasopharyngeal isthmus.
3. Defective smell: Sensation becomes dulled due to constant irritation of the nasal mucous membrane by the regurgitated food.
4. Defective hearing: Due to oedema of the orifice of the auditory tube resulting in a blockage consequent upon pharyngeal inflammation from the regurgitated food.
5. Repeated respiratory tract infection.
6. Chance of aspiration bronchopneumonia.
7. Defective dentition because of irregular development of alveolus.
8. Cosmetically ugly look particularly when associated with cleft lip.

Surgical anatomy of nasopharyngeal isthmus

It is the site of communication between oral and nasal parts of the pharynx. It presents the following boundaries:

- *In front:* Posterior surface of the soft palate.
- *Behind:* Passavant's ridge which is a slightly curved transverse ridge close to the arch of atlas and contains palatopharyngeus sphincter muscle.
- *On each side:* Palatopharyngeal arch.

Isthmus is closed during deglutition or blowing of air through the mouth and in expression of some elements of speech (except M and N) by the following muscles:

- (i) Levator palati which elevate the soft palate.
- (ii) Tensor palati which stretch the soft palate, and
- (iii) Contraction of palatopharyngeal sphincter

which approximates the Passavant's ridge with the soft palate.

Treatment of cleft lip**Operation**

Aim of operation: Mainly for cosmetic reason and also for functional recovery.

Optimum time for operation: The operation on cleft lip should be performed within 6 months from birth i.e., *before the primary dentition*. Otherwise, the defective dentition is difficult to correct. Moreover, protruded teeth may cause pressure on the suture line and prevent wound healing.

A 'Rule of 10' (Millard) is usually advocated which indicates that the baby should fulfill the following criteria:

- (i) the age of the baby should be 10 weeks or more;
- (ii) the weight of the baby should be 10 lbs;
- (iii) the haemoglobin content should be 10 gms.

So, the operation is better performed when the baby is 2 1/2 months to 3 months old.

Reasons for this timing:

- (a) Nutrition of the baby is acceptable for general anaesthesia and operation.
- (b) As the lip elements are larger, the repair is technically more precise and easier.
- (c) Dropper feeding in post-operative period is easier, thereby facilitating healing of the lip wound by reducing the need for suckling with the freshly wounded tissues.

Some authorities advise operation within 48 hours of birth, if the baby is otherwise healthy. They offer the following arguments:

- (a) There is no undue risk in doing such early operation because the child is prepared for birth trauma as the child is provided with maternal antibody.
- (b) Such early operation facilitates sucking which provides great comfort to the mother.

But arguments against such early operations are:

- (i) The lip is very thin and tiny; so, repair is often inaccurate, necessitating a second operation later on.
- (ii) Since this is not an emergency condition and since the deformity will not increase significantly by waiting for 10 weeks, no harm is done.

Advantages of early operation:

1. Better final results are likely.
2. It facilitates effective sucking.
3. It helps in moulding of the alveolus. When cleft lip is associated with cleft alveolus, early operation induces reduction or even closure of the alveolar gap.
4. It avoids defective speech.
5. It allays worries of the parents.
6. When cleft lip is associated with cleft palate, early reconstruction of the lip reduces the gap in the palate to some extent.

Principles of operation:

1. The cleft lip should be so repaired that the normal shape and symmetry of the lip is maintained. For this, adequate mobilisation of the flaps from the underlying maxillae on both the sides of the cleft is necessary. It also avoids tension on the suture line.
2. To gain adequate vertical length from the nostril floor to the vermilion border to match precisely with the contralateral side of the lip. To achieve this various methods (Millard, Mirault-Blair, Le Mesurier, Tennison, etc.) have been proposed which are variations of a Z-plasty. *Millard's technique is popular and its principle is 'rotation down and advancement closure' of the lip elements.*
3. Paring of the edges so that there may be precise union.
4. Accurate apposition of the margins: To achieve this suture should be done in three separate layers—
 - (i) Mucous membrane to mucous membrane – by catgut.
 - (ii) Muscle to muscle – by catgut or vicryl.
 - (iii) Skin to skin – by silk or vicryl.

Essentials of operation:

1. Pouting of the lip – The upper lip must be placed well ahead of the lower.
2. The vermilion border of lip (i.e. the mucocutaneous junction) should be intact; no portion of the mucosa should be everted over the skin.
3. Cupid's bow should be intact.
4. Integrity of the orbicularis oris should be maintained by proper suturing of muscle layer.

5. Associated nostril deformity should be corrected. Minor deformity can be corrected at primary operation but a major one necessitates a second operation at a later age while the nasal growth is stopped.

Pre-operative measures:

1. Mouth cavity should be free of infection. For this throat swab may be necessary for culture and antibiogram.
2. Antibiotic spray in the mouth to eradicate any infection.
3. The baby should be habituated with spoon-feeding or dropper-feeding.

Post-operative measures:

1. Application of Logan's bow at the conclusion of operation as an immediate post-operative measure may help. Advantage is two fold:
 - (i) the child cannot scratch over the suture line;
 - (ii) it may also relieve tension over the suture line.
2. The arms and hands should be tied on some suitable splint.
3. Maintenance of nutrition by liquid diet – spoon-feeding or dropper-feeding.
4. Antibiotic – Both systemic and local spray in the mouth.
5. The stitches are removed on the 5th or 6th day after operation.

When the child will be allowed to suck the mother's breast?

The normal tensile strength of a scar is regained in fourteen days. Hence, though the stitches are removed on the fifth or sixth day, the baby should not be allowed to suck the mother's breast before the third week.

Treatment of cleft palate**Operation: Palatoplasty**

Aim of operation: To achieve mainly adequate speech. For this both length and mobility of the palate are necessary.

Optimum time for operation: 14 to 18 months i.e., before the child speaks, because once the child has learnt to speak it is very difficult to correct nasal intonation. Moreover, the child is reasonably stable to tolerate blood loss during operation.

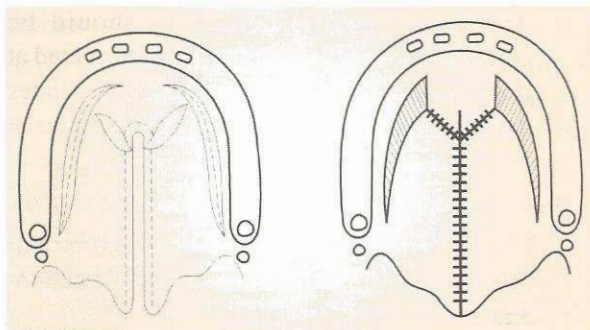


Fig. 19.18. Wardill's method of operation.

But modern trend is to repair the palate a bit earlier i.e., between the age of 9 months and 1 year because a child starts making efforts to produce understandable sound as early as that.

Some authorities suggest that early repair of the cleft of hard palate may cause maldevelopment of the maxillae which will be evident in later life. So, according to them, the ideal treatment is early repair of the soft palate only before the child begins to speak, to allow its development as an essential organ of speech. Closure of the hard palate defect should be postponed until the facial development is well advanced i.e., till the time of secondary dentition. Till that time the defect in the hard palate can be closed either with an obturator or dental prosthesis.

Reasons for this timing:

1. Before the age of nine months the tissues are delicate and friable and so are difficult to handle. There is a chance of failure of operation.
2. The cleft palate baby suffers from malnutrition which should be corrected.
3. There is a chance of spontaneous reduction of the gap in the hard palate, which may occur up to the age of 1 years. But spontaneous reduction occurs by dropping or even collapse in limited cases of the palatine vault, which may result in maxillary deformity in later age. For this, waiting for spontaneous reduction is not preferred by some groups of surgeons.

Principles of operation:

1. To repair the cleft in both the soft and hard palates.
2. To create a mobile soft palate of sufficient length to allow closure of the nasopharyngeal

isthmus which is necessary to achieve adequate speech without any nasal intonation and also to prevent regurgitation of food in the nasopharynx.

Essentials of operation:

1. Cleft should be repaired in two layers:
 - (i) Muco-periosteal flaps on the nasal side in the first layer, and then,
 - (ii) Muco-periosteal flaps on the oral side.
2. For mobilisation of the flaps on the oral side, lateral longitudinal release incisions should be made on either side at the junction of palate and alveolus (Fig. 19.18).
3. For adequate mobilisation of the posterior part of the flaps, the pterygoid hamulus is to be broken off on many occasions to release the tensor palati muscles.
4. Greater palatine vessels should be preserved. Otherwise there will be flap necrosis.
5. For adequate closure of the nasopharyngeal isthmus some form of pharyngoplasty may be necessary.

Method of operation: Various methods have been proposed to achieve complete closure of the cleft in the hard palate. Most of these are based on the original Langenbeck procedure.

Fig. 19.18 illustrates Wardill's "Four-flaps" method of repair which combines the repair of the cleft with elongation of the soft palate.

Cleft palate closure can be achieved by one- or two-stage palatoplasty also. In the first stage – the soft palate cleft is closed followed by closure of hard palate at the second stage. The philosophy of two-stage closure encourages a physiological narrowing of the hard palate cleft to minimise surgical dissection at the time of the second procedure.

Pre-operative measures:

1. Criteria for suitability of operation:
 - (i) The child must weigh about 20 lbs.
 - (ii) Haemoglobin content should be 12 gms percent.
2. Normal B.T. and C.T.
3. Mouth cavity should be free of infection.
4. The child should be made habituated with spoon-feeding or dropper-feeding.
5. 150-175 cc of blood should be kept ready for transfusion during operation, if necessary.

Post-operative measures:

1. Maintenance of nutrition by liquid diet – spoon-feeding or dropper-feeding.
2. Antibiotic – both systemic and local spray in the mouth.
3. Intensive speech training.

Pharyngoplasty

It is the reconstruction of the pharynx where the nasopharyngeal isthmus is made narrowed by plastic surgical procedures. This will prevent nasal escape of air during speech and thus avoid nasal intonation. There are several techniques:

1. *Wardill's pharyngoplasty*: Transverse incision is made on the posterior pharyngeal wall at the level of Passavant's ridge – followed by vertical suture.
2. *Denis-Browne pharyngoplication*: Purse-string suture of the mucosa is applied over the posterior pharyngeal wall at the level of Passavant's ridge.

3. *Pharyngeal flap technique* – either:

- (i) inferiorly based posterior pharyngeal flap – to bridge a smaller gap, or
- (ii) superiorly based posterior pharyngeal flap – to bridge a larger gap.

Note: Many children with cleft lip and palate require orthodontic treatment to correct the dental problems.

CONGENITAL ANOMALIES OF URETHRA

HYPOSPADIAS

Hypospadias is a common congenital anomaly of the urethra where the external urinary meatus is situated on the undersurface (ventral surface) of penis or perineum, instead of its normal position at the tip of the glans. In most cases, there would be a ventral bending of penis known as *chordee*.

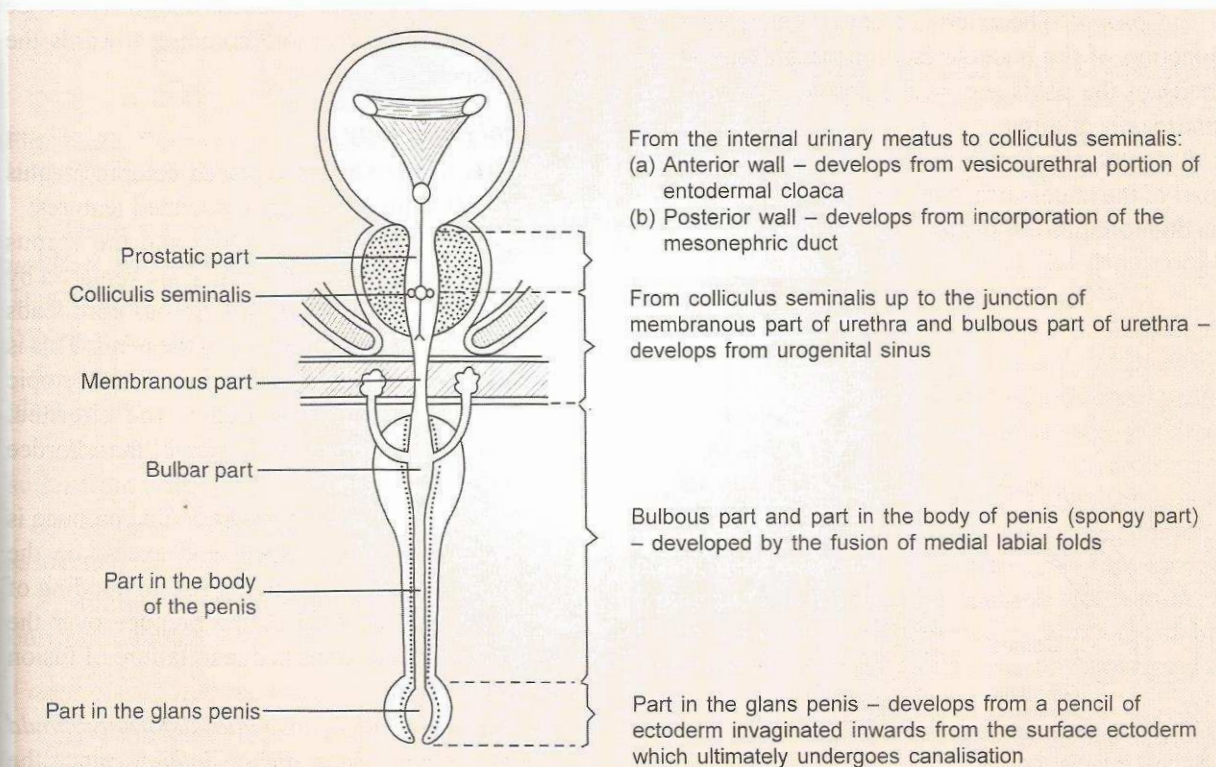
DEVELOPMENT OF URETHRA

Fig. 19.19. Different parts of the urethra and their development.

Incidence

It occurs about 1 in 350 births.

Types of hypospadias (Fig. 19.20)

Anatomically classified according to the site of external meatus.

Site	Type	Percent
Glans penis	Glandular	80 percent
Corona	Coronal	
Shaft of the penis	Penile	
Junction of penis and scrotum	Penoscrotal	20 percent
Median raphe of scrotum	Scrotal	
Perineum	Perineal	

1. **Glandular:** Commonest and less severe and is due to failure of development of ectodermal urethral groove. External meatus is situated on the undersurface of the glans penis with a blind depression at the normal site. There is no chordee. Glans is flattened and the penis is straight and rarely bent. This variety does not require any treatment *except a meatotomy if the ectopic orifice is too small*.

2. **Coronal:** The external meatus is situated at the junction of the undersurface of the glans with the body of the penis (i.e. at the corona glandis). The chordee is minimum.

3. **Penile:** The external meatus is situated at some part of the undersurface of the body of the penis and is due to failure of fusion of the anterior part of the

two medial labial folds. *Chordee is a prominent feature and the penis becomes curved ventrally.*

4. **Penoscrotal:** The meatus opens at the junction of the penis with the scrotum.

5. **Scrotal:** The meatus is situated anywhere along the median raphe of the scrotum. The scrotum may not be bifid completely but is very often diminutive. Sex differentiation is usually not a problem.

6. **Perineal:** The scrotum is completely bifid and the urethra opens either between its two halves or anywhere along the perineal raphe. The penis is usually quite small with marked chordee.

This type is very often associated with bilateral maldescended testes in which event the sex differentiation may be difficult.

Both the scrotal and perineal types are due to complete failure of fusion of genital folds after the rupture of urogenital membrane.

Anterior groups of hypospadias (i.e. glandular, coronal, penile) are more common than the posterior groups (i.e., scrotal, perineal); *why?* – because the fusion of the medial labial folds begins in the posterior aspect and then proceeds anteriorly. Hence deficiency of fusion is more common towards the anterior aspect.

Surgical pathology

In addition to the ventrally placed ectopic meatus there are following important associated features:

1. **Chordee:** The absent urethra and the corpus spongiosum distal to ectopic orifice are replaced by fibrous cord. Contracture of this fibrous cord leads to downward (ventral) curvature of the penis. This is known as chordee. The more proximal is the ectopic meatus, the more pronounced is the chordee. Whenever there is erection of penis, the chordee becomes more prominent.

2. **An incomplete ventral prepuce:** The prepuce is deficient on the ventral aspect and excess on the dorsal aspect of the glans, giving rise to hooding of the prepuce. The absence of the prepuce over the ventral aspect of the glans is due to failure of fusion of the genital folds.

Any child born with hypospadias deformity should not have a circumcision during infancy, since the redundant skin (prepuce) over the dorsal aspect will prove useful later during the reconstruction of urethra.

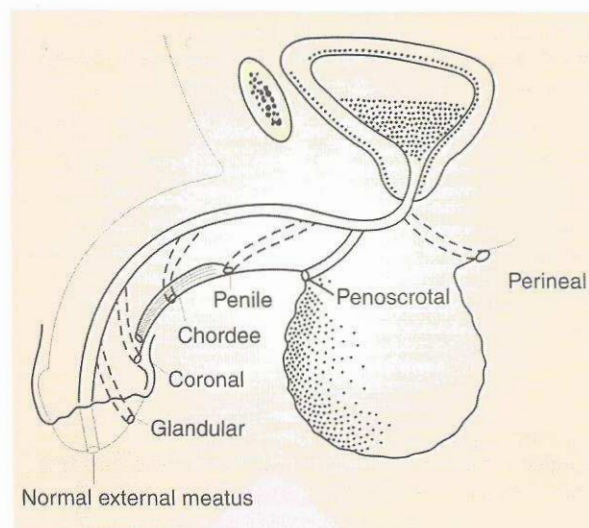


Fig. 19.20. Different types of hypospadias.

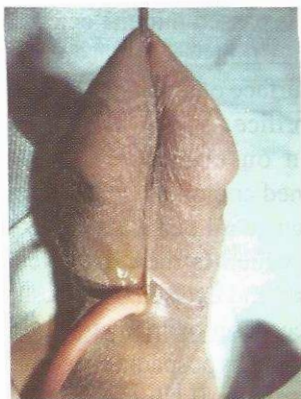


Fig. 19.21. Distal penile hypospadias. (Courtesy: Dr. Uday Shankar Chatterjee, MS, MCh (Paed), MCh (Uro))



Fig. 19.22. Penoscrotal hypospadias. (Courtesy: Dr. Uday Shankar Chatterjee, MS, MCh (Paed), MCh (Uro))

3. There may be *stenosis of the ectopic external urethral orifice* causing difficulty in passing urine. This is more common with glandular and coronal types.

4. There may be a small blind pit on the tip of the glans penis.

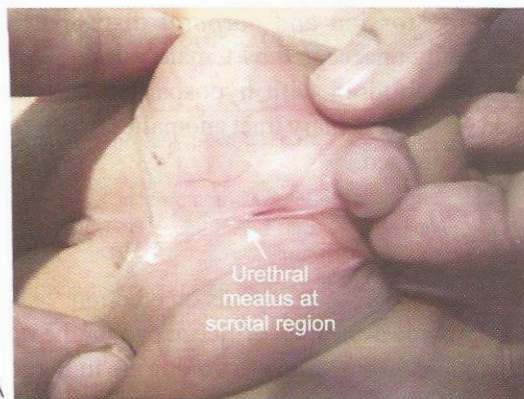
5. The child with hypospadias usually does not suffer from urinary incontinence.

6. The perineal or scrotal type is very often associated with bilateral undescended testes.

7. Multiple urethral orifices may be present, but only one communicates with the urethra per se.

Problem with hypospadias

1. Hypospadias results in an abnormal urinary stream. The stream of the urine is deflected due to downward deformity of the glans penis resulting in spoiling of the undergarments.



A



B

Fig. 19.23. (A) Scrotal hypospadias. (B) Bifid scrotum also noted in the same patient. Scrotal hypospadias is usually associated with bifid scrotum. (Courtesy: Dr. Uday Shankar Chatterjee, MS, MCh (Paed), MCh (Uro))

2. Infertility is frequently associated with a proximal penile, scrotal, perineal and penoscrotal meatus.

3. Hypospadias with ectopic orifice in the scrotal or in the perineal area is very often accompanied by bilateral maldescended testes with quite a small penis in which event one should suspect 'intersex' problem and investigate accordingly. They must be differentiated from adrenogenital syndrome and pseudohermaphroditism. Before performing urethroplasty, determination of the genetic sex by karyotyping and adrenal function test by determination of 17 oxysteroids and 17 ketosteroids should be performed. Laparotomy or laparoscopy and gonadal biopsy may be required.

4. Chordee is exaggerated during erection and may cause painful erection.

5. Due to chordee, the intercourse may be impossible or uncomfortable.

6. Penile torsion and corporal disproportion: Cosmetically unacceptable by female partner.

7. Some children with hypospadias may have associated upper urinary tract anomalies which need to be investigated.

Treatment

Objectives of treatment

1. To produce a straight penis on erection.
2. To provide a terminal orifice at or near the tip of the glans, from which to void urine in the long axis of the penis in standing position.
3. To make possible insemination well into the vagina.
4. To achieve a good cosmetic or aesthetic appearance.
5. To achieve a urethra free of meatal stenosis, stricture, fistula, sacculum.

Glandular hypospadias usually does not require any treatment, but may require meatotomy and dilatation of the orifice, if it is small or stenosed.

All other types are treated by some plastic reconstructive procedures (urethroplasty). Several plastic operations are available including two-stage procedures like Denis-Browne's, Byars, Cecil-Culp operations. Multiplicity of the procedures signifies that no single operation is universally satisfactory or free from complications.

In recent years the trend has been towards one-stage rather than two-stage procedures and towards repairs that give a good cosmetic as well as functional results.

Timing of operation

The operation is preferably completed before the age of 5 years i.e., before the school-going age.

DENIS-BROWNE'S TECHNIQUE

It is a two-stage operation and is described here because of its simplicity.

Stage I: Chordee correction to cause straightening of the penis – It is performed preferably between 1 and 2 years.

Stage II: Construction of the urethra – It is performed preferably between 4 and 6 years.

Stage I: Chordee correction

1. The child is placed in supine position.

2. A probe is passed into the ectopic orifice.
3. Incision: A transverse incision is made on the ventral surface of the penis 1.25 cm distal to the ectopic orifice.
4. The skin on either side of the urethra is undermined and mobilised.
5. The fibrous cord distal to the ectopic orifice is exposed which is then dissected from before backwards and excised. As a result, the ectopic meatus recedes proximally towards the perineum i.e., a coronal hypospadias becomes a penile one or a distal penile hypospadias becomes a proximal penile one.
6. The skin flaps are closed longitudinally, and if necessary, to relieve tension a release incision may be given in the midline on the skin of the dorsum of the penis.

Stage II: Construction of the urethra

Principle of reconstruction

The urethral tube is made by burying a strip of skin along the line of urinary meatus which then grows out at its edge and rolls itself into a tube within 8 to 10 days. This reconstructive procedure is always combined with urinary diversion from the seat of operation either by:

- (a) perineal urethrostomy, or by
- (b) suprapubic cystostomy particularly in case of penoscrotal type.

The idea of urinary diversion is to avoid urinary leakage and soiling of the reconstructed urethra.

Technique

Step I: Perineal Urethrostomy (Fig. 19.24A) or Suprapubic Cystostomy.

Step II:

1. A 'U' shaped incision is made on the under surface of the penis. The incision embraces the ectopic orifice proximally (Fig. 19.24A).
2. The skin flaps are undermined and mobilised both in lateral directions and in the direction of perineum beyond the old urethral opening. When the posterior undermining involves the scrotum, two stab wounds are made on either side of the scrotum to prevent haematoma (Fig. 19.24B).
3. A triangular area of skin is raised from the glans on either side of the proposed new meatus.

Different other plastic repairs of hypospadias now available according to the site of meatus

Site of meatus	Recommended methods
Glandular	Usually does not require any plastic repair, except meatotomy if the meatus is stenosed.
Coronal or sub-coronal	MAGPI (Meatal Advancement and Glanduloplasty Integrated).
Distal penile (when the meatus is on the distal shaft approximately 1 cm from the corona)	Mathieu procedure (Perimeatal based flap) — one-stage procedure.
Proximal penile (with a proximal meatus with or without chordee)	Vascularised preputial island flap mobilised to the ventrum either by Asopa or Duckett technique — one-stage procedure.

- The mobilised skin flaps are apposed and sutured in the mid-line preferably with 'double-stop' sutures of monofilament nylon. The first stop is the sterilised glass beads and the second stop is the sterilised small sections of soft aluminium tubing, which are crushed and thus hold the sutures in place (Fig. 19.24C).
- A release incision is made on the skin along the whole length of the dorsum of the penis to relieve tension. The release incision prevents necrosis of the flaps and thus urethral fistula.

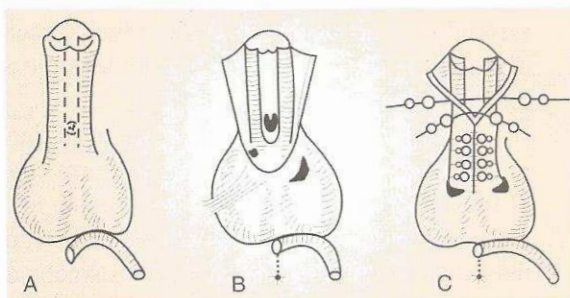


Fig. 19.24. Steps of Denis-Browne's operation.

EPISPADIAS

It is the congenital anomaly where the external urethral opening is situated on the dorsal aspect of the penis instead of its normal situation at the tip of the glans.

Incidence

It is a rare condition occurring about 1 in 30,000 births.

Types

Three types (Fig. 19.25)

- Glandular.
- Penile.
- Totalis — where the urethra is deficient throughout its whole dorsal extent. This variety

is very often associated with the ectopia vesicae (Fig. 19.26A&B).

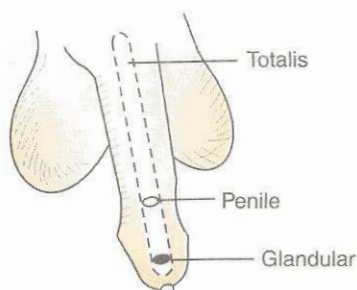


Fig. 19.25. Types of epispadias.

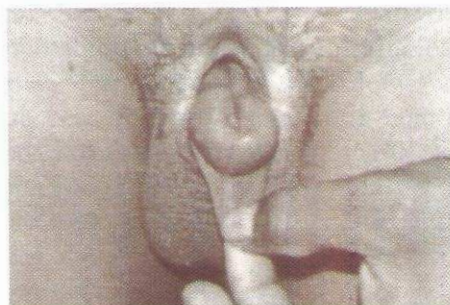


Fig. 19.26 A. Epispadias totalis. Note the hooded skin (pulled down) on the ventral aspect of the glans penis.



Fig. 19.26 B. Epispadias totalis.

Treatment

Glandular type usually does not require any treatment.

For the other two varieties operation is required. A urethroplasty, similar in principle of Denis-Browne's technique, is undertaken usually at the age of 3 years.

ECTOPIA VESICAE

(Syn. Exstrophy of the bladder) (Fig. 19.27A&B)

It is a congenital anomaly characterised by the absence of the infra-umbilical part of the anterior abdominal wall and the anterior wall of the urinary bladder.

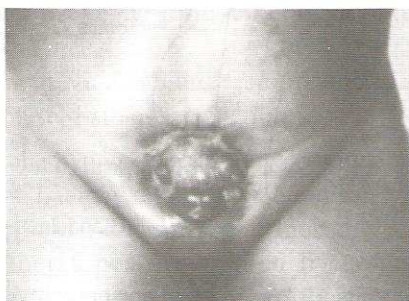


Fig. 19.27A. Ectopia vesicae in female. Umbilicus was absent.

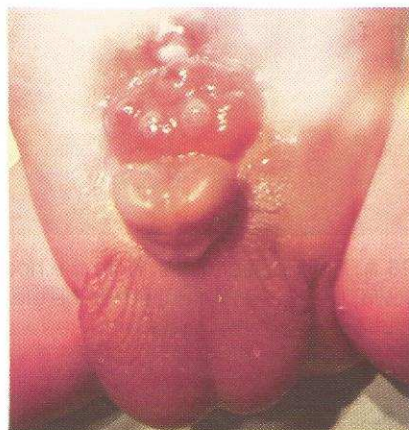


Fig. 19.27B. Ectopia vesicae in male. Note the glans penis and the scrotum.

Developmental background

The defect is due to failure of migration of mesodermal cells along the genital folds from the primitive streak to the infra-umbilical part of the anterior abdominal wall. At the same time the phallus grows from the caudal end of the urogenital membrane.

Therefore, when the elongated urogenital membrane ruptures, the interior of the bladder and part of the urethra are exposed to the surface.

Incidence

It is a rare condition occurring about 1 in 50,000 live births.

Diagnostic criteria

1. This condition is two to three times more common in boys.
2. Red congested mucosa covering the posterior wall of the bladder is exposed in the lower part of the anterior abdominal wall. This deep red posterior bladder wall may protrude out through the defect due to the pressure of viscera behind it.
3. When the exposed mucous membrane is drawn upwards, the wet trigone is visible and also effluxes of urine from the ureteric orifices can be seen.
4. The exposed mucous membrane bleeds readily on touch.
5. A well-defined line of demarcation between the protruding mucous membrane and the adjacent abdominal skin (i.e., the mucocutaneous junction) can be delineated.
6. If the protruding bladder wall is reduced deep to the muco-cutaneous junction, the firm edge of the defect in the abdominal wall can be felt.
7. Constant dribbling of urine with excoriation of the surrounding skin and continuous smells of urine.
8. There may be the following associated abnormalities:
 - (a) Wide separation of the symphysis pubis, and due to this defect, the child usually walks with waddling gait. *X-ray will confirm the gap.*
 - (b) Absence of the umbilicus.
 - (c) Umbilical hernia.
 - (d) Bilateral inguinal herniae with undescended testes (when sex differentiation is difficult).
 - (e) Epispadias totalis: The penis is broader and shorter than the normal. The penis is often in two separate portions.
 - (f) Bifid clitoris in girls. There may be a wide separation of the labia minora exposing the vaginal orifice.

- (g) Anterior placement of the anal orifice with or without rectal prolapse.
- (h) Rarely ureteric dilatation: *IVU assessment of upper urinary tract is preliminary to surgery.*
- (i) Occasionally extra-urogenital anomalies e.g., atresia of oesophagus or duodenum.

Complications

1. Painful ulceration of the exposed urothelium.
2. Squamous metaplasia of the exposed urothelium which may become polypoid in later years.
3. Neoplastic change of the exposed urothelium due to long continued irritation: Adenocarcinoma which is locally malignant and only rarely metastasizes.
4. Recurrent attacks of ascending pyelonephritis.
5. 50 percent of the patients die because of renal failure before the age of thirty.
6. Continuous bad odour of urine which makes the patient a nuisance to his relatives.
7. Cosmetically unsightly.

Treatment

Parental distress and inevitable complication demands some form of management policy in these patients. Surgical treatment is either reconstructive or diversionary. Prognosis appears to be better in girls.

A. Bladder reconstruction

To be performed within first three years of life.

1. Reconstruction of the bladder wall after mobilisation.
2. Preliminary iliac osteotomy to allow approximation of symphysis pubis.
3. Closure of the anterior abdominal wall.
4. In the male, repair of epispadias to be performed at the age of six years.
5. Sphincter control is difficult to achieve because of absent sphincter muscles and nerve supply;

thus the reconstructed bladder becomes an incontinent reservoir. Recently, electronically controlled continent device is implanted in bladder neck musculature to achieve continence.

Advantages:

The operation

- (i) removes the unsightly exstrophied bladder and gives a satisfactory external appearance.
- (ii) keeps the patient reasonably dry. The girls may need intermittent self-catheterisation whereas boys may require condom drainage.
- (iii) may preserve the genital function in males.

But very soon the operation becomes complicated with ascending pyelonephritis due to the persistence of vesico-ureteric reflux. Moreover, there is a chance of stone formation and there may be overflow incontinence. So, in future a urinary diversion will be necessary.

B. Urinary diversion

Excision of the urinary bladder is combined with urinary diversion either into the colon (uretero-sigmoidostomy) or into an ileal or sigmoid loop conduit.

Ureterosigmoidostomy is contraindicated in presence of anal prolapse with defective anal musculature. Moreover, it carries a high risk of development of chronic renal failure due to ascending infection within 20 years of operation. So, ileal or sigmoid loop conduit becomes the operation of choice.

Hazards of urinary diversion:

1. Fistula at the site of anastomosis.
2. Stenosis at the site of anastomosis resulting in bilateral hydronephroses.
3. Recurrent ascending pyelonephritis.
4. Hyperchloraemic acidosis with hypopotassemia – more in case of ureterocolic anastomosis.
5. Renal failure.

20

CHAPTER



Soft Tissue Sarcoma

Soft tissue sarcomas are uncommon malignant neoplasms arising from the soft tissues of connective tissue origin i.e., mesodermal in origin.

- **Mesodermal:** Connective tissue origin: sarcoma
 - Soft tissue sarcoma
 - Bone sarcoma
- **Ectodermal origin:** A neuroectodermal contribution corresponding to the peripheral nerves are also included in soft tissue sarcoma.
- **Endodermal:** Epithelial tissue origin: carcinoma.

SOFT TISSUE SARCOMA

Age

- Accounts for less than one percent of all malignancies diagnosed in adults.
- Relatively common in younger population (c.f. carcinoma common in elderly population).
- Accounts for 15% percent of all malignancies diagnosed in children under the age of 15 years.
- Ranked fifth in cancer related malignancy incidence and death in children under the age of 15 years.
- Common varieties noted in different age group:
 - *Children* – embryonal rhabdomyosarcoma
 - *Young adult* – synovial cell sarcoma
 - *Older adult* – liposarcoma or malignant fibrous histiocytoma (MFH)

- Some of the pediatric lesions are congenital.
- *Interestingly it is noted that congenital soft tissue sarcomas rarely behave malignantly though they show an aggressive behavior in their microscopic appearance.*

Sites

May develop in any anatomic sites:

- 50% in lower extremity
- 30% in trunk
- 20% in upper limb and head & neck.

Common histopathology according to anatomic site

- *Extremity:* Liposarcoma, malignant fibrous histiocytoma
- *Retroperitoneal or visceral:* Liposarcoma, leiomyosarcoma
- *Abdominal wall:* Desmoid tumour

Aetiology

- Little is known about aetiopathogenesis
- Genetic Predisposition
 - (i) von Recklinghausen's disease—neurofibrosarcoma
 - (ii) Li-Frument syndrome
 - (iii) Retinoblastoma
 - (iv) Gardner's syndrome (Familial adenosis polyposis)

- Lymphoedema
 - (a) Post-surgical e.g., lymphangiosarcoma may develop from post mastectomy lymphoedema of upper limb.
 - (b) Post-irradiation
 - (c) Parasitic infestation (Filariasis).
- Ionising radiation
- Trauma:
 - (i) Post-parturition—may give rise to abdominal wall tumour e.g., Desmoid tumour
 - (ii) Extremity—it is uncertain whether trauma induces malignant change or trauma draws attention to already existing lesion.
- Chemical:
 - (i) Polyvinyl chloride
 - (ii) Thorium dioxide
 - (iii) Arsenic
 - (iv) Haemochromatosis.

It is to be noted that:

- With the ubiquitous presence of connective tissue, soft tissue sarcoma may develop anywhere in the body; *somatic site is more common than visceral site. Extremity tumours are predominant.*
- Cell of origin is of minimal value for prognostication and treatment schedule.
- Dermatofibrosarcoma protuberans is a low grade neoplasm.

- Alveolar rhabdomyosarcomas are high grade neoplasms.
- Sarcomatous cells may reproduce tissue similar to that from which it arises, e.g., liposarcoma, fibrosarcoma, osteosarcoma or chondrosarcoma.
- As in other field of oncology, attempts are being made to classify soft tissue tumours on the basis of their gene expression profile.

Origin

- *De-novo*: Large majority of soft tissue sarcomas arise de-novo.
- *Secondary*:
 - (i) Malignant change may occur in pre-existing benign tumours e.g., lipoma, fibroma, neurofibroma.
 - (ii) May arise as a complication of radiotherapy e.g. M.F.H.

Prognosis

Based on following criteria:

1. *Cell of origin*: It is of little value for prognostication and treatment schedule.
2. *Histological type*: One of the most important predictors of sarcoma specific death.
Malignant peripheral nerve sheath tumour has the highest risk of mortality.

Pathological Classification (according to histogenic concept)

Cells of origin	Benign lesions	Malignant lesions	Locally malignant lesions
Adipose tissue (Fat cell origin)	Lipoma	Liposarcoma	
Fibrous tissue (Fibroblast origin)	Fibroma	Fibrosarcoma (low grade malignancy) Malignant fibrous histiocytoma (high grade malignancy)	Extra-abdominal desmoid
Nerve tissue (Schwann cells origin)	Neurilemoma Neurofibroma	Neurofibrosarcoma	
Striated muscle	Rhabdomyoma	Rhabdomyosarcoma	
Smooth muscle	Leiomyoma	Leiomyosarcoma	
Lymph vessels	Lymphangioma	Lymphangiosarcoma	
Blood vessels	Haemangioma	Haemangiosarcoma	
Synovial membrane	Ganglion Synovioma (Giant cell tumour)	Synovial sarcoma	
Mesothelium	Mesothelioma	Malignant mesothelioma	
Histiocytes	Dermatofibroma	MFH	Dermatofibrosarcoma protuberans

SARCOMA



Fig. 20.1. Liposarcoma right thigh. History of soft tissue swelling in the upper part of right thigh for many years. The swelling suddenly started to increase in size during last six months with appearance of pain and stiffness more during walking. The lump was firm with ill-defined margin but became hard and less mobile on contraction of quadriceps muscles – the diagnosis was (?) sarcoma. Total excision biopsy of the lump including 2 cm of surrounding tissue around the lump in continuity was performed. Histopathology confirmed liposarcoma. Patient was referred to oncologist for radiotherapy.



Fig. 20.3. Neurofibrosarcoma of the right upper arm – note multiple subcutaneous nodules over the face, trunk, and right upper limb which were present since childhood. Sarcomatous changes developed on the right arm nodules which were rapidly growing during last 8 months. (Courtesy: Prof. Bansari Goswami, MS)



Fig. 20.2. Fibrosarcoma outer side of middle of the right leg. (Courtesy: Prof. Diptendra Sarkar, MS, DNB, FRCS)



Fig. 20.4. Synovial sarcoma right knee. (Courtesy: Prof. Sudeb Saha, MS, FAMS)



Fig. 20.5. Dermatofibroma left groin – note the cystic degenerative nodules on the surface. (Courtesy: Prof. Sudeb Saha, MS, FAMS)

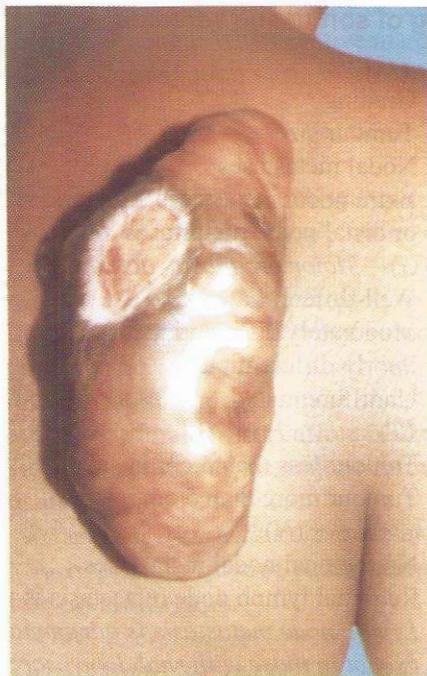


Fig. 20.6. Dermatofibrosarcoma over the upper left back (shoulder) – firm to hard irregular lump. Note the ulcer on the surface with pale granulation tissue. (Courtesy: Prof. Sudeb Saha, MS, FAMS)

3. Size:

- Less than 5 cm in size is associated with good prognosis regardless of histological grade. Chances of metastasis is low if appropriately excised at the time of initial treatment.
- Large size is associated with poor grading and poor prognosis.
- Large size is associated with dedifferentiation, so poor grading.
- Large size is associated with late recurrences.

4. Depth of lesion:

Superficial lesions do better than the deep lesions. With deep lesions chances of local invasion along the fascial planes, vessels and nerves are more common.

5. Grade:

- *Most important prognostic factor.*
- Based on several factors as follows:
 - (i) Cellularity – hypocellular or hypercellular.
 - (ii) Differentiation – well-differentiated or poorly differentiated.

- (iii) Degree of mitoses – less than 10 mitoses or more than 10 mitoses per high power field.
- (iv) Stromal content—very little stroma or abundant stroma.
- (v) Necrosis—absent or present.
 - Low grade tumour—low risk of subsequent metastasis (15 percent).
 - High grade tumour—high risk of subsequent metastasis (50 percent).

6. Presence or absence of metastasis:

(a) Metastasis by blood spread is common.

- Sarcomas often grows rapidly, and dissemination occurs early by blood stream.
- Pulmonary metastasis are common with extremity lesion.
- Extra pulmonary metastasis is rare, and usually found only with advanced and disseminated disease.
- Liver is frequent site of metastasis with retroperitoneal or visceral sarcomas.

(b) Nodal metastases: Only occasional tumour metastasises by lymphatics are:

- (i) Rhabdomyosarcoma
- (ii) Synovial sarcoma

7. Surgical excision:

The most important prognostic factors for survival being completeness of resection and grade of the tumour. (See Treatments.)

Diagnosis

A. Extremity lesion:

- Usually presents as asymptomatic, slowly growing soft tissue lump.
- Generally painless lump
- Suspicious lesions are:
 - (i) Enlarging lump
 - (ii) Appearance of pain in a painless lump
 - (iii) Lump greater than 5 cm in size
 - (iv) Any new lump that persists beyond 4 weeks.

B. Intra-abdominal or retroperitoneal lesion:

- Non-specific abdominal pain or discomfort or gastrointestinal symptoms.
- Abdominal palpation detects the lump.

Investigation

A. To establish the diagnosis:

For tissue diagnosis and grading – **Biopsy**

1. Tissue biopsy of the soft tissue lump is must, specially to know the grade of the tumour.
 2. *Types of biopsy in extremity lump:*
 - (a) FNAC – Grading is not possible
 - (b) Core biopsy – Grading may be difficult
 - (c) Incisional biopsy—
 - (i) Provides adequate sample, so grading is possible.
 - (ii) Advocated for large extremity lump.
 - (iii) The incision should be longitudinal and be centered over the lump in its most superficial location so that the entire biopsy tract can be included in subsequent radical treatment whether surgical or irradiation.
 - (d) Excisional biopsy
 - (i) Recommended for small cutaneous or subcutaneous lump between 3 and 5 cm in size.
 - (ii) Grading is possible.
 3. *Types of biopsy in retroperitoneal lump or intra-abdominal lump.*
 - (a) FNAC or CT guided core biopsy has a limited role.
 - (b) May be indicated if abdominal lymphoma is strongly suspected.
 - (c) In most patients, exploratory laparotomy and resection of the tumour settle the diagnosis.
- ### B. To know the extent of the disease:
- (a) Chest x-ray – for lung metastases.
 - (b) Abdominal CT, chest CT
 - (c) MRI – Procedure of choice. MRI enhances the contrast between the lesion and the adjacent structures. It also helps to determine the relationship of the lesion to nearby blood vessels and nerves.
 - (d) Liver should be imaged as a part of abdominal CT or MRI.
 - (e) Angiography – May be of value in delineating tumour-vessel relationship and hence the treatment.
 - (f) Lymphangiography – *only when rhabdomyosarcoma or synovial sarcoma is suspected as these sarcomas only may metastasize by lymphatics.*

Staging of soft tissue sarcoma

Based on GTNM

- G** Grading
T Tumour size
N Nodal metastases
M Extra nodal metastases e.g., lung and liver, or distal nodal metastases

Grade (G)—Major prognostic determinate

- G1 Well-differentiated
 G2 Moderately differentiated
 G3 Poorly differentiated
 G4 Undifferentiated

Tumour (T)

- T1 Tumour less than 5 cm in size
 T2 Tumour more than 5 cm in size.

Nodal metastases (N)

- N0 No regional nodal metastases
 N1 Regional lymph node metastasis is present.
Lymph node metastasis is uncommon except in those with rhabdomyosarcoma and synovial sarcoma.

Metastases (M)

- M0 No distant metastasis
 M1 Distant metastasis present.
 • Lung metastasis is common with extremity lesions.
 • Liver metastasis is common with retroperitoneal lesions.

GTNM staging system

Stage I	G1	T1 - T2	NOMO
Stage II	G2	T1 - T2	NOMO
Stage III	G3	T1 - T2	NOMO

NO: No lymph node metastasis.

MO: No distant metastasis.

Stage IV	G1 - G3	T1 - T2	NIMO
	G1 - G3	T1 - T2	NIMI

Lymph node metastasis ± Distant metastasis.

TREATMENT

Surgery

Extremity lesion

Aim is

- (i) to achieve local control, and
- (ii) to treat metastases

Surgical treatment

- (i) Wide surgical excision is the first line of treatment for all soft tissue sarcoma.

- (ii) The surgical objective should be complete removal of tumour with negative margins and maximal preservation of function.

Types of surgery

1. *Excisional biopsy:*
 - (i) All gross tumour is excised *but tumour remains behind because of pseudoencapsulation.*
 - (ii) *Recurrence rate is 90 percent.*
2. **Wide excision with 1 to 2 cm of surrounding normal tissue in continuity** is advocated to prevent local spread but at the same time to provide maximal preservation of function. Deliberate sacrifice of the major neurovascular structures should be avoided.
 - (i) *Recurrence after wide excision is 50 percent.*
 - (ii) **So requires adjuvant radiotherapy and/or chemotherapy.**
 - (iii) *Wide excision followed by adjuvant irradiation of tumour bed has resulted in the achievement of limb sparing surgery without compromise of survival.*
3. **Radical excision implies compartmental excision:**
 - (i) *Recurrence rate is 20 percent.*
 - (ii) Requires adjuvant radiotherapy and/or chemotherapy for low recurrence rates.
4. **Amputation at suitable level i.e., one joint above the lesion or disarticulation.**
 - (i) Most radical surgery.
 - (ii) Recurrence rate is 5 percent.
 - (iii) Little advantage over limb sparing surgery.
 - (iv) May be advocated in below knee lesions or in upper limb lesions after reasoning to both patient and patient party.
5. **Prophylactic lymph node resection is not indicated in soft tissue sarcoma.**

Role of radiotherapy

Aim is to reduce local recurrence.

Radiotherapy can be given as

- (i) curative
- (ii) post-operative adjuvant
- (iii) pre-operative neoadjuvant.

Curative radiotherapy

Radiotherapy has little effect on either the primary growth or on the secondary growth.

Note that sarcoma of bone is sensitive to radiotherapy and so can be used as an alternative to amputation.

(a) Indication:

- Poor surgical risk patients
- Patient refusing surgery
- Surgery not feasible because of anatomical location e.g., retroperitoneal tumour.
- Tumour > 5 cm in size

(b) Type of radiation:

- (i) Sophisticated radiation like megavoltage or linear accelerator.
- (ii) High dose radiation (7000 to 8000 rads).

(c) Results of curative radiotherapy versus combined modality treatment i.e., Surgery + radiotherapy.

- Recurrence rate is higher with only radiotherapy.
- Moreover delayed fibrosis is common with only radiotherapy

Post-operative adjuvant radiotherapy

(a) Indication:

- Extremity lesion who have undergone surgery less than amputation.
- Truncal sarcoma who have undergone removal of gross tumour only.
- Retroperitoneal tumour where total removal of tumour is not possible.
- Lesions less than 5 cm do not benefit much from radiotherapy due to excellent general prognosis following surgery.

(b) Type of radiation:

- Brachytherapy for high grade lesions only.
- External beam radiation therapy for high grade or low grade lesion.

Preoperative neoadjuvant radiotherapy

- Used in locally advanced disease.
- Little value.

Role of chemotherapy

- As an adjuvant therapy its role is not established.
- May be used as a part of definitive treatment of metastatic disease.
- May benefit in large and high grade lesions.
- May benefit for recurrent lesions.
- Combination chemotherapy is used.

- **Leiomyosarcoma** shows greater resistance to chemotherapeutic drugs.
- Synovial sarcoma shows good response to ifosfamide based chemotherapeutic drugs.
- Doxorubicin (adriamycin), ifosfamide, dacarbazine, cyclophosphamide, DTIC and mensa have been used alone or in combination.
 - (i) VAC – Vincristine, Adriamycin, Cyclophosphamide.
 - (ii) DVC – Doxorubicin, Vincristine, Cyclophosphamide.
 - (iii) CYVADTIC – Cyclophosphamide, Vincristine, Adriamycin, DTIC.

Doxorubicin (Adriamycin) is the single most effective drug but best used with various other drug combinations.

Result of chemotherapy

- (i) Length of survival is improved (Median survival > 1 year).
- (ii) Percent of patients live at 5 year is increased.

Neo-adjuvant chemotherapy with or without radiation therapy

- (i) May cause downstaging of the tumour.
- (ii) So, may be used in large primary lesions.
- (iii) May be used for limb sparing procedures in difficult cases.

FIBROSARCOMA

- Origin from mesenchymal cells and composed of spindle cells.
- May occur wherever there is fibrous tissue.
- Common sites are the trunk and the limbs, particularly the inner and intra-muscular fibrous septae of adductors, scapulohumeral and pectoral muscles.
- May occur at any age.
- It may arise as a localised, firm, mobile tumour which eventually gets fixed.
- Usually slowly growing but eventually pulmonary metastases appear.
- Tumour is radioresistant.
- Treatment is by wide local excision or amputation depending on the site of the lesion, supported by radiotherapy.

SYNOVIAL SARCOMA

- Synovial sarcoma is highly malignant.
- Predominant cells are spindle shaped.
- Can originate from the synovial tissue in joints, tendon sheaths or bursae.
- Spreads widely, infiltrating its surroundings including bone.
- Metastasize both by lymphatics and blood.
- Tumour is usually radioresistant.
- Treatment is by wide local excision or amputation depending on the site of the lesion, supported by radiotherapy.
- Shows good response to ifosfamide-based combination chemotherapy.

MALIGNANT FIBROUS HISTIOCYTOMA (MFH)

- Origin from histiocytes and primitive mesenchymal cells have been proposed.
- Aggressive soft tissue sarcoma
- *Occurs principally as a mass at the extremities (70%); more on the lower limbs.*
- May occur in the abdominal cavity, retroperitoneum, head and neck, trunk and in other parts of the body.
- Situated in the deep tissues of the extremities and trunk. It involves deep fascia or skeletal muscles. Rarely confines to subcutis without fascial involvement.
- *Occurring principally in middle-aged individuals.* Peak incidence is in the fifth decade.
- Rare symptoms and signs include episodic hypoglycemia and rapid tumour enlargement during pregnancy.
- **Diagnosis:** FNAB
- **Treatment:** Wide margin of excision of tumour is advocated.
Pre- or postoperative radiotherapy is also given to the lesion larger than 5 cm in diameter.

RETROPERITONEAL NEOPLASM

Boundaries of retroperitoneal space

It is a potential space between the peritoneal cavity and the posterior abdominal wall. It is bounded:

- *Above* – by diaphragm
- *Below* – by pelvic levator ani muscle
- *Anteriorly* – by posterior parietal peritoneum

- *Posteriorly* – by vertebral column with posterior abdominal muscles on either side - psoas, quadratus lumborum and transversus abdominal muscles.

Pathology

Common benign tumours

1. Lipoma
2. Lymphatic or chylous cysts
3. Cysts of urogenital origin
4. Enterogenous cysts

Common malignant tumours

1. Lymphoma
2. Lymphosarcoma
3. Liposarcoma
4. Fibrosarcoma
5. Leomyosarcoma
6. Rhabdomyosarcoma
7. Neuroblastoma

Retroperitoneal malignancies

Malignant tumours in the retroperitoneal space may result from:

- (a) Primary neoplasm in the retroperitoneal organ such as the kidney, adrenal, pancreas and colon. *These are excluded from the domain of retroperitoneal tumours.* These are organ specific tumour.
- (b) Metastases from a remote primary malignancy into the retroperitoneal lymph nodes.
- (c) Primary neoplasm of the retroperitoneal lymphatic system, e.g., lymphoma.
- (d) Primary neoplasm of the soft tissues of the retroperitoneum of mesodermal origin such as adipose tissue, fibrous tissue, smooth and striated muscle, blood vessels, lymphatic tissues and nerve tissues.
- (e) Primary germ cell neoplasm arising from embryonic rest cells.

The most common primary malignancy of the retroperitoneum is sarcoma.

RETROPERITONEAL SARCOMA

Primary malignant tumours of retroperitoneum arising from the soft tissues of mesenchymal or mesodermal origin occupying the potential retroperitoneal

space such as adipose tissue, connective tissue, smooth and striated muscles, blood vessels, lymphatic tissue and nerve tissue – known as retroperitoneal sarcoma.

- Incidence is 0.3 to 3 percent.
- *Sarcomas are solid*, hard and usually fixed mass (cf. *cystic tumours are predominantly benign*).
- Grow slowly and silently and present as asymptomatic abdominal lump when they become quite large.
- Sarcomas may present with following symptoms:
 - (i) Vague abdominal pain or heaviness, anorexia, weight loss.
 - (ii) Obstructive gut symptoms – vomiting, altered bowel habits, intestinal obstruction.
 - (iii) Urinary tract symptoms – haematuria, dysuria, oliguria/anuria.
 - (iv) Neural compression symptoms leading to radicular pain, paraesthesia
 - (v) Vascular and/or lymphatic compression leading to limb swelling and heaviness.
 - (vi) With or without any of these symptoms, abdominal palpation may detect the large intra-abdominal mass accidentally.
- They are locally aggressive but rarely metastasize.
- Local recurrence after complete resection is common.
- Patients with high grade sarcoma have a higher risk of systemic disease and death.
- *Associated with a poor long term survival rate, main reason being the extreme difficulty encountered in performing a complete surgical excision with a rim of normal tissue around the tumour because of tumour adherence to adjacent major vascular structures and nerves.*
- 5 years survival rate is 25% even after radio- and chemotherapy.

Investigation

1. CT/MRI

- (a) Non invasive
- (b) Most effective means of delineating abnormalities of retroperitoneum.
- (c) Provide information—
 - (i) Regarding exact site, location and extent of the tumour, physical characteristics – solid or cystic. *Cystic lesions are predominantly benign.*

(ii) Tumour relationship to major vascular structures.

(iii) Presence or absence of metastasis.

2. Biopsy

- (a) US or CT guided FNAC or core biopsy has limited role.
- (b) *Helps to detect the tissue of origin. Grading of tumour is not possible.*
- (c) Usually reserved for those lesions with strong suspicion of lymphoma, or germ cell tumour.
- (d) In most patients, exploratory laparotomy and resection of the tumour settle the diagnosis.

Treatment

1. Surgery
2. Radiotherapy
3. Chemotherapy

Surgery

- (a) Complete enbloc resection of the tumour with tumour free margin is the best from of treatment.
- (b) Any adjacent involved organ should also be resected e.g. intestine, kidney.
- (c) Parietal fixation does not contraindicate resection

(d) Only contraindications are

- (i) Wide spread metastases
- (ii) Lymphoma.

Lymphoma is chemosensitive and radiosensitive tumour and can be diagnosed preoperatively by US or CT guided core biopsy and so surgery is avoided.

Radiotherapy

Indications of radiotherapy:

1. Radiosensitive tumours e.g., lymphomas
2. All inoperable tumours
3. Residual tumour following incomplete resection
4. As an adjunct to surgery for some tumours
 - (a) Neuroblastoma
 - (b) Liposarcoma
 - (c) Rhabdomyosarcoma
5. Tumour recurrence

Chemotherapy

Indications of chemotherapy:

1. Lymphomas
2. Leomyosarcoma showed greater resistance to radiotherapy.
3. In other sarcomas with metastases—may be used as a part of definitive treatment.

21

CHAPTER



Intestinal Stomas

An intestinal stoma is an artificial opening of the intestine, small or large, onto the abdominal wall to divert the faeces and flatus to the exterior, where they may be collected in a changeable bag. Common intestinal stomas are colostomy and ileostomy.

COLOSTOMY

A colostomy is a surgical procedure where an artificial opening is constructed between a part of the colon and the anterior abdominal wall to allow the discharge of faeces.

- A colostomy is designed to be either temporary or permanent.
- Colostomy has no sphincteric control.

Types of colostomy

1. According to anatomic location
2. According to function
3. According to appearance
4. According to duration

According to anatomic location

- End sigmoid colostomy – Left iliac fossa
- End descending colon colostomy – Left iliac fossa
- Transverse colostomy – Above & right to the umbilicus
- Caecostomy – Right iliac fossa

The appropriate stoma site must be selected preoperatively for all the elective procedures and for most emergent operations.

According to function that the colostomy is intended for

- Diverting colostomy or defunctioning colostomy.
- Decompression colostomy

A. Diverting colostomy

Any colostomy may be regarded as 'Diverting' if it is so constructed that faeces are prevented from entering the diseased bowel distal to it. There are two types: temporary and permanent.

- (a) *Temporary diverting colostomy*: Usually loop colostomy – the purpose is to provide temporary diversion of faeces for:
 - (i) Protection of the complicated and threatened distal anastomosis e.g., following anterior resection for rectal cancer.
 - (ii) Grossly infected diverticulitis with or without perforation.
 - (iii) Traumatic injuries of the colon or rectum
 - (iv) Multiple, complicated and high perianal fistulae.

Once the distal lesions are under control, the temporary colostomy is closed.

- (b) *Temporary diverting end colostomy*: Hartmann's procedure.
- (c) *Permanent diverting colostomy*: An end colostomy – the purpose is to provide permanent diversion of faeces for:

- (i) Perforated unresectable rectal cancer.
- (ii) Late and unresectable rectal or anal cancer.
- (iii) Unreparable lesion of rectum or anal canal following trauma, Crohn's disease, hidradenitis suppurativa.

B. Decompression colostomy – Temporary

- The purpose is to provide decompression of hugely dilated large bowel proximal to obstructing growth of the rectum or sigmoid colon.
- Frequently done on emergency basis to prevent impending rupture of the dilated colon proximal to obstruction.
- Useful and life-saving.
- Provides opportunity for a subsequent definitive cancer surgery without compromise of the principles of cancer surgery.

Types of decompression colostomy

- A loop transverse colostomy
- 'Blow hole' stoma constructed in the caecum or transverse colon.
- Tube type caecostomy

According to appearance of colostomy

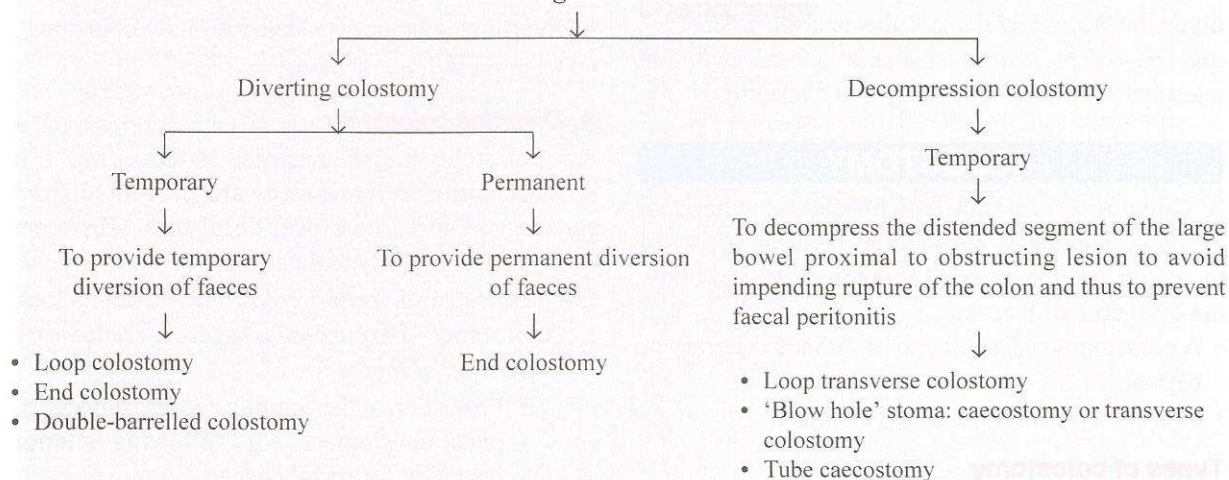
- (i) End colostomy
- (ii) Loop colostomy
- (iii) Double-barreled colostomy
- (iv) 'Blow hole' transverse colostomy
- (v) 'Blow hole' caecostomy
- (vi) 'Tube' caecostomy

According to Duration

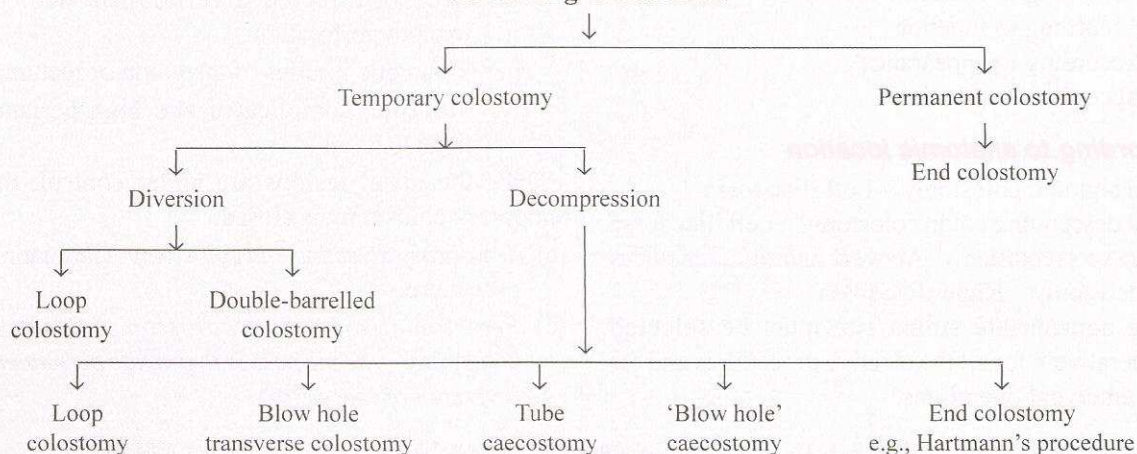
Temporary colostomy

- (i) Loop colostomy

According to Function



According to Duration



- (ii) End colostomy
- (iii) Double-barrelled colostomy

Permanent colostomy

- End colostomy

TEMPORARY COLOSTOMY

- **Loop colostomy:** Commonly done with the transverse colon or sigmoid colon.
- **Double-barrelled colostomy:** Commonly done with sigmoid colon or transverse colon.

Idea: To divert the faecal stream temporarily to defunction the distal colon.

Indications

1. To relieve a distal obstruction of the sigmoid colon due to any cause, commonly carcinoma or diverticulitis.
 - Usually performed as an emergency procedure to relieve obstruction as it is quick and easy to make.
 - The resection of the diseased part is carried out later on in a completely defunctioned segment of the colon which is empty, inactive and relatively sterile.
2. To defunction the distal colon and thus to protect a distal anastomosis after a low colorectal anastomosis.
3. To prevent faecal peritonitis after perforation of the colon following obstructive cancer, diverticulitis colon, traumatic injuries of colon or rectum
4. To facilitate the operative repair of high fistula in ano.
5. To prevent spoiling of the urinary bladder in vesico-colic fistula or vagina in vagino-colic fistula.

Types

A. Temporary loop colostomy

Method:

1. A temporary loop colostomy is made by bringing a loop of bowel to the surface of abdomen.
2. The loop is held in place by a glass or plastic rod or a rubber tube passed through the mesentery.
- When a glass or plastic rod is used, a piece of rubber tubing is attached to both ends to prevent the rod from slipping out.

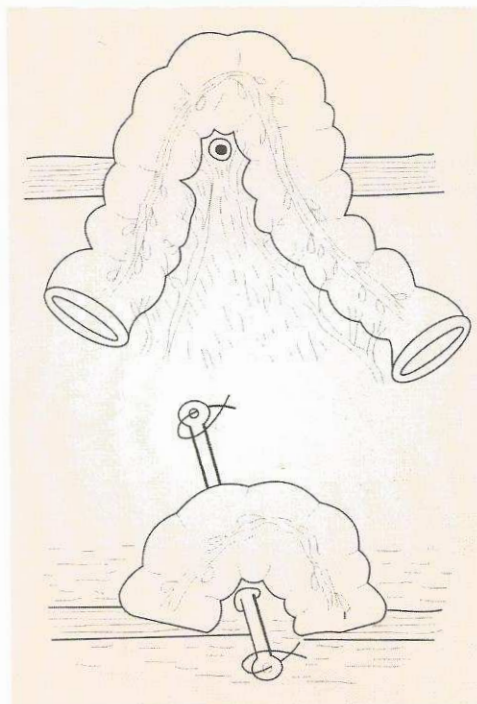


Fig. 21.1. Loop colostomy. Glass or plastic rod or thick rubber tube cut from the malecot catheter is inserted through the mesentery to support the loop and sutured to the abdominal wall.

- If a rubber tube is used, it is stitched to skin beyond the stoma.
3. So, loop colostomy is made by using the large bowel that has long mesentery e.g., transverse colon, sigmoid colon.
 4. Most loop colostomies are made in the transverse colon in diseases involving the left side of the colon.
 5. *Site of election:*
 - (a) Transverse colostomy is usually brought out above and to the right side of the umbilicus.
 - (b) Sigmoid colostomy is usually brought out in the left iliac fossa.
 6. Proximal half of the transverse colon to the right of the middle colic artery is the preferred site, so that the entire left colon being left available for the subsequent formal operations in the future.
 7. The colonic loop so brought out is sutured to the layers of the abdominal wall to prevent retraction and/or faecal soiling of the peritoneal cavity.

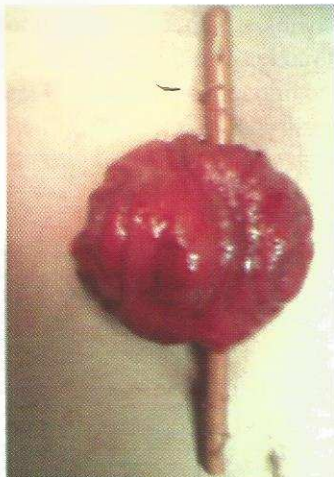


Fig. 21.2. Temporary transverse loop colostomy held in place by a rubber tube.

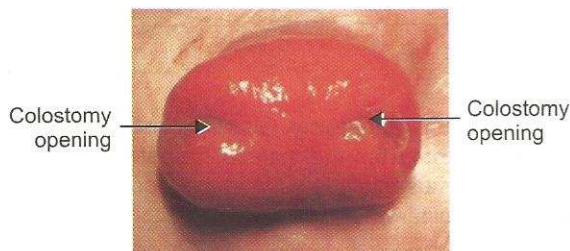


Fig. 21.3. Mature transverse colostomy with healthy moist pink stomas.

8. The colostomy is opened along its exposed tenia coli to minimise bleeding from gut wall.
9. The opening may be made:
 - (a) *Immediately* – when colostomy is done specially for distal obstruction i.e., for decompression
 - (b) *After 48 hours* – when colostomy is made for other indications.
10. The glass or plastic rod is removed on the tenth postoperative day and replaced by the rubber tubing, which is finally removed in 2 to 3 weeks.
11. *Closure of colostomy:*
 - (a) Temporary colostomy is closed following the surgical cure of the distal lesion for which the temporary stoma has been made.
 - (b) Colostomy closure is safely and easily accomplished once the stoma is mature i.e., usually 2 months after the colostomy has been established.

(c) Intraperitoneal closure of colostomy is preferred over extraperitoneal closure to avoid closure breakdown resulting in fresh faecal fistula.

B. Double-barrelled colostomy (Devine or Paul-Mickulicz procedure)

Type of defunctioning temporary colostomy

- A variant of temporary diverting proximal colostomy with mucous fistula (cf. proximal ileostomy with mucous fistula).
- Can be performed with the lesion of sigmoid or transverse colon which has a long mesocolon.
- Rarely used now-a-days in poor risk patients, e.g. after resection of a gangrenous sigmoid volvulus, grossly infected diverticulitis or malignant growth of the sigmoid colon with or without perforation.
- *Two types:* Devine procedure and Paul Mickulicz procedure.

Devine procedure (Fig. 21.4)

- A variant of temporary double-barrelled colostomy.
- Occasionally performed with the transverse or sigmoid colon *which has a long mesocolon*.
- After resection of the intervening diseased part of the colon intraperitoneally, both the proximal and distal colon ends are brought out to the exterior through two separate incisions (openings) on the abdominal surface thereby fashioning a skin bridge between the stomas. This skin bridge prevents the spillover of faeces from proximal colostomy opening to distal colostomy opening.

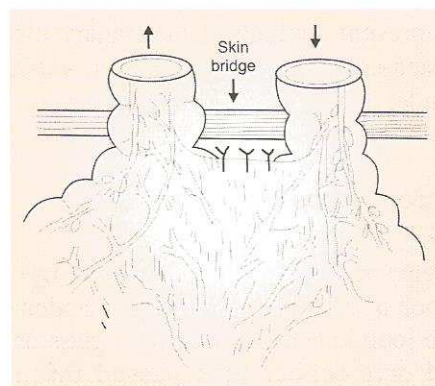


Fig. 21.4. Devine procedure. Skin bridge between the stomas will prevent spillover of faeces from proximal stoma to distal stoma. Requires intraperitoneal closure of colostomy openings when the purpose is served.

- Once the purpose is served, a formal *intra-peritoneal closure of colostomies* is required usually after 6-8 weeks.

Paul-Mickulicz procedure (Fig. 21.5)

- A variant of temporary double-barrelled colostomy.
- Occasionally performed for tumours of the sigmoid colon or transverse colon which has a long mesocolon.
- Performed in poor risk patients who are too old and frail to withstand a major resection and anastomosis at the same sitting.
- Also performed in an emergency situation for obstruction due to growth of the sigmoid colon when no bowel preparation is possible.
- The tumour is mobilised and exteriorised through a *separate but same muscle cutting incision* in the abdominal wall, e.g., left iliac fossa for sigmoid growth.
- The tumour is then resected extraperitoneally.
- The adjacent walls of the two limbs of the colon below the colostomy openings are sewn together at the antimesenteric borders to form a spur keeping the ends of the guts in contact (*spur colostomy*).
- Once the purpose is served, the colostomy openings are closed *extraperitoneally* after crushing the intervening spur by an *enterotome* to restore the bowel continuity.

Note: For tumours of the sigmoid colon in a poor risk patient, Hartmann's procedure may also be performed.

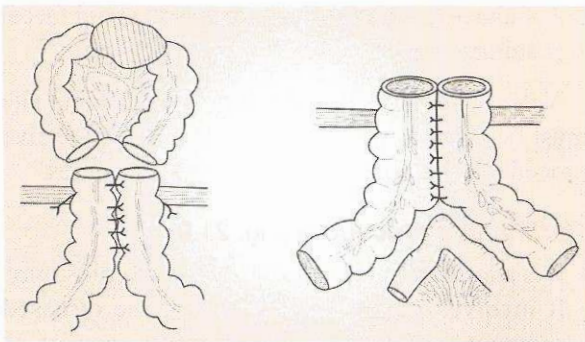


Fig. 21.5. Paul Mickulicz procedure. After the purpose is served, the intervening spur between colon ends are crushed by the crushing enterotome and the colostomy openings are closed extraperitoneally.

PERMANENT COLOSTOMY (END COLOSTOMY)

Types

- **End sigmoid colostomy:** Most common type.
- **End descending colon colostomy:** When the inferior mesenteric artery is transected during an operation of rectal cancer.

Idea: To provide a permanent opening to allow excretion of faeces for the rest of life in absence of anus.

Indications

Usually performed after abdominoperineal resection (APR) for lower 1/3rd, middle 1/3rd, and upper 1/3rd of rectal cancer or anal cancer: APR + left iliac colostomy is commonly done.

(Note: In upper 1/3rd rectal cancer, anterior resection can be performed in selected cases by experienced surgeon.)

1. **Method:** It is formed by bringing the distal open end of the proximal colon after excision of the distal colon along with rectum and anal canal.
2. **Site of election:**
 - (a) Commonly left iliac fossa
 - (b) The stoma location should be flat and away from the skin creases and the bony prominences of costal or iliac margins. It should be in an area of normal healthy-appearing skin that is visible to the patient. This avoids the faecal spillage from the bag onto the abdominal surface.

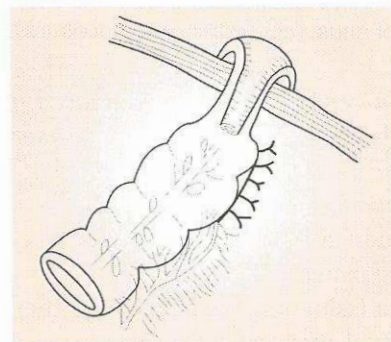


Fig. 21.6. End colostomy. Colostomy opening everted and attached to skin margin of the hole. Sutures closing the lateral peritoneal space to prevent small intestine's herniation.



Fig. 21.7. Left iliac permanent end colostomy in an elderly patient after abdominoperineal resection for lower 1/3rd of rectal carcinoma. Note the healthy moist pink stoma projecting well above the abdominal surface like a nipple. Examine the patient's perineum for presence of scar and absence of anus. (cf. Hartmann's procedure). (Courtesy: Dr. Sudeb Saha, MS, FAMS)



Fig. 21.8A. Permanent left iliac colostomy covered with colostomy bag (following APR for lower third rectal cancer). Always examine the perineum for scar and absence of anus. (cf. Hartmann's procedure)

In general, the stoma is placed just above the spino-umbilical line at the lateral border of the rectus abdominis muscle or at the middle of the spino-umbilical line. In obese individuals, the stoma may be placed higher, so patient can see it.

3. While fashioning the colostomy, after bringing the distal open end of the colon to the abdominal surface, special care is taken to close the lateral space between the intraperitoneal segment of the colon and the peritoneum of the lateral abdominal wall. *This is to prevent internal herniation of small bowel through the defect.*



Fig. 21.8B. Same patient after removal of bag. Note that the colostomy was healthy and projecting well from the skin surface but appeared narrow – so, developed stenosis of the stoma. However, patient was managing well with the stenosed stoma with frequent finger dilatation.

Note also the hypertrophy of the lower half of the midline scar; but no evidence of incisional hernia on straining or coughing.

4. Alternatively, a *retroperitoneal tunnel* is developed to bring the distal open end of the colon to the surface to avoid closure of the lateral space (Goligher).
5. Some prefer to bring the colostomy through the midline abdominal incision or medial belly of left rectus muscle. There is no need to close the lateral spaces, as these are so large that movements of the small bowel can take place freely without any obstruction.
6. The stoma should protrude 2–5 cm – *like a nipple* – above the skin surface. This is to avoid faecal spillage under the bag.

Note: When possible, the appropriate stoma site must be selected preoperatively for all the elective procedures and for most emergency procedures.

Hartmann's procedure (Fig. 21.9)

- It is a form of temporary diverting end colostomy.
- It involves segmental excision of the diseased segment (either from diverticulitis, cancer or gangrene) + closure of the distal stump + temporary end colostomy of the proximal colon.
- This procedure requires a second major laparotomy for reanastomosis of the divided ends of the bowels at a later date, if necessary.

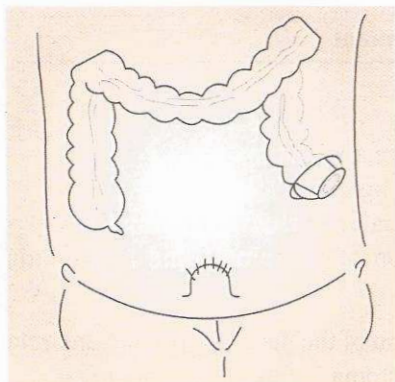


Fig. 21.9. Hartmann's procedure (see text) with left iliac end colostomy + sutured intrapelvic rectal stump (cf. left iliac end colostomy following APR). *Examine the perineum for the presence of anus and digital examination for presence of rectal stump. In APR there will be presence of scar in the perineum and absence of anus.*

Indications

1. Grossly infected diverticulitis with or without perforation of the sigmoid colon.
2. Obstructing lesion from cancer of the sigmoid colon or upper 1/3rd of rectum.

Colostomy care

1. Colostomy usually starts functioning during the first 5-7 days following operation. If colostomy fails to function by the 7th postoperative day, straight X-ray abdomen must be done to rule out small bowel obstruction.
2. Look that collecting bag with or without flange or stoma adhesive disc are fitting properly around the stoma. Patient should personally learn and manage to apply the bag properly. Patient may require nurse's assistance during early postoperative period.
3. Patient should take proper care to keep parastomal area clean and dry. Patient should ensure good local hygiene and apply properly fitting collecting bag to avoid parastomal cellulitis.
4. In the presence of parastomal cellulitis, patient requires repeated local application of antiseptic solution and spirit around the stoma and the bag.
5. Patient should avoid diet which produces loose stool. Patient should take high fibre diet and plenty of drinks to avoid constipation.
6. Patient should learn self-finger dilatation of the stoma to prevent stomal stenosis.

7. Patient may require frequent colostomy irrigation with 1 litre of plain water through an enema tube for the purpose of dilating the proximal colon sufficiently to produce a reflex peristaltic contraction of the gut that evacuates the entire distal colon.

Patient and nurse should take extreme caution while passing the enema tube to avoid perforation of the colon proximal to the stoma which will cause severe abdominal pain during irrigation. This complication represents a surgical emergency and requires urgent laparotomy and reconstruction of colostomy.

CAECOSTOMY

Indications

Severely ill patient with massive distension of large bowel with impending rupture of caecum. The massive distension may be due to:

- (i) Growth distal to caecum particularly when ileo-caecal valve is competent leading to closed loop obstruction.
- (ii) Pseudo-obstructive syndrome, e.g., ileus in elderly patient.

It provides immediate and very effective drainage both of the colon and of the lower ileum.

Purpose

- Caecostomy is performed to prevent faecal peritonitis from rupture of caecum.
- Colonoscopic decompression is a better alternative for cases of pseudo obstruction.
- Caecostomy is performed only when the other methods of decompression fail.

Types

Two types:

1. Skin sutured caecostomy or 'Blowhole' caecostomy.
 2. Simple tube caecostomy.
- A caecostomy is only a temporary measure to allow a few days to improve the general condition of the patient following decompression of large bowel.
 - Definitive surgery is carried out after 5 to 7 days of establishing the caecostomy.
 - The tube caecostomy requires repeated irrigation with normal saline to prevent it from being blocked by the faecal matter.
 - In tube caecostomy, the tube may be removed after the 10th postoperative day if it is no longer required.

Complications of colostomy

Management

Immediate

- | | |
|---|--|
| <ol style="list-style-type: none"> 1. Bleeding from edges <ul style="list-style-type: none"> – <i>Immediate after operation</i> – <i>Delayed</i>, usually from granuloma around the margin of colostomy 2. Necrosis and sloughing of the distal end of the stoma | <ul style="list-style-type: none"> • Pressure application. • Suture ligation. • Chemical cauterisation of granuloma. If persistent, excision of granuloma and refashioning of the stoma. • Excision of the necrosed portion and refashioning of the stoma. |
|---|--|

Intermediate

- | | |
|--|---|
| <ol style="list-style-type: none"> 1. Retraction (Fig. 21.10) 2. Prolapse (Fig. 21.11) 3. Stenosis (Fig. 21.8A&B) 4. Fistula 5. Abscess 6. Stricture | <ul style="list-style-type: none"> • Colostomy refashioning. • The protruded bowel should be covered with hypertonic saline packs and then gradually and gently pushed back. If recurrent, colostomy refashioning. • Finger dilatation. If failed, refashioning of the stoma by splitting the constricting abdominal wall layers. • Colostomy refashioning. |
|--|---|
- Common with Crohn's disease
7. *Parastomal cellulitis*
- Poor construction or location of the stoma leads to maceration and inflammation of the skin, pseudoepitheliomatous hyperplasia at the mucocutaneous junction of the stoma
- | | |
|--|--|
| <ol style="list-style-type: none"> 8. Intraperitoneal abscess | <ul style="list-style-type: none"> • Avoid loose stool. • Ensure good local hygiene. • Apply proper fitting bag. • Drainage with or without antibiotics. |
|--|--|

Late

- | | |
|--|---|
| <ol style="list-style-type: none"> 1. Colostomy hernia 2. Colostomy diarrhoea <ul style="list-style-type: none"> – Spurious diarrhoea – Infective enterocolitis 3. Recurrence of malignancy at the colostomy margin. | <ul style="list-style-type: none"> • Closure of the lateral space and refashioning of colostomy. • Avoid constipation. • Avoid food which causes constipation and/or diarrhoea. • Responds to oral neomycin or metronidazole. • Poor prognosis. Excision of distal bowel segment including stoma and reestablishment of colostomy at a fresh site. |
|--|---|



Fig. 21.10. Retraction of colostomy stoma causing spillage of stool onto the abdominal surface.

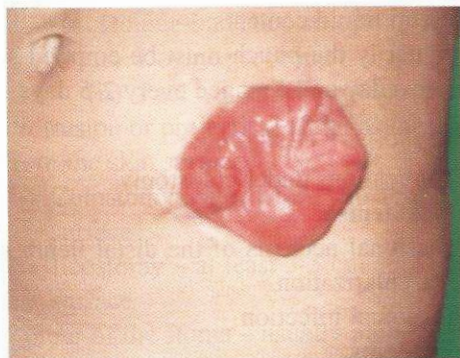


Fig. 21.11. Prolapse of the mucosa through the left iliac colostomy stoma.

ILEOSTOMY

Ileostomy is a surgical procedure where an opening is constructed between the distal ileum and the abdominal wall.

- Ileostomy has no sphincteric control.
- Ileostomy is designed to be either temporary or permanent.

Indications

Ileostomy is necessary when disease or injury has rendered the large bowel incapable of safely processing the intestinal waste typically because the colon has been partially or totally removed.

Diseases of large bowel which may require surgical removal of the colon include:

- Inflammatory bowel disease—ulcerative colitis, Crohn's disease.
- Familial adenomatous polyposis.
- Total colonic Hirschprung's disease.
- Occasionally in the treatment of colorectal cancer where the tumour is causing blockage.

Types

Two types:

- Permanent ileostomy or end ileostomy
- Temporary ileostomy
 - (i) End ileostomy
 - (ii) Loop ileostomy

Indications

(a) **Permanent end ileostomy:** It is performed in conjunction with a total proctocolectomy in inflammatory bowel diseases like Crohn's disease, ulcerative colitis or polyposis coli.

(b) **Temporary loop ileostomy:**

- It is performed when temporary diversion of ileal contents is required.
- Commonly performed to divert the ileal contents temporarily to protect a tenuous ileorectal or ileoanal anastomosis following total colectomy or proctocolectomy, e.g., adenomatous polyposis.

Advantage: To allow healing of the distal anastomosis well and thus allow safe passage of faeces through the anal orifice in future after closure of ileostomy.

(c) **Temporary end ileostomy with mucous fistula:** Occasionally a temporary end ileostomy of the proximal end of the ileum and mucous fistula of the distal end of the ileum are constructed, after resection of a gangrenous segment of bowel or a perforated caecal lesion, when primary anastomosis is contraindicated.

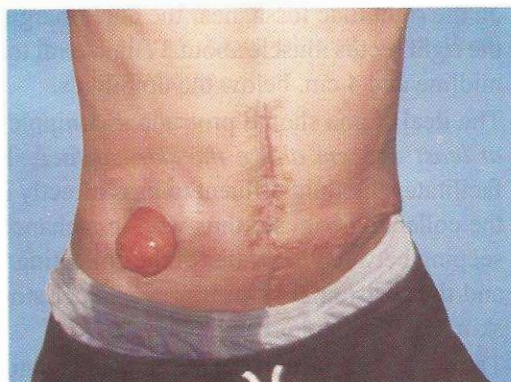


Fig. 21.12. Right iliac permanent end ileostomy – note the stoma was healthy, moist and pink and projecting well above the abdominal surface like a nipple.

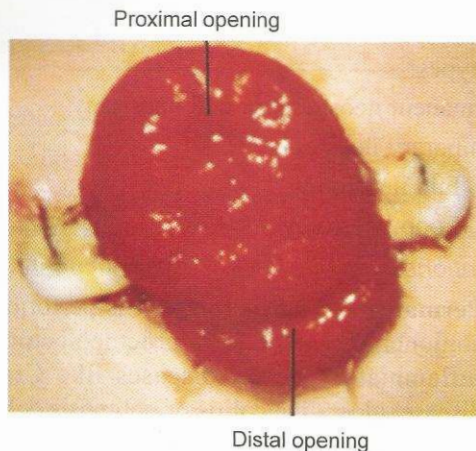


Fig. 21.13. Right iliac temporary loop ileostomy held in place by a plastic rod.

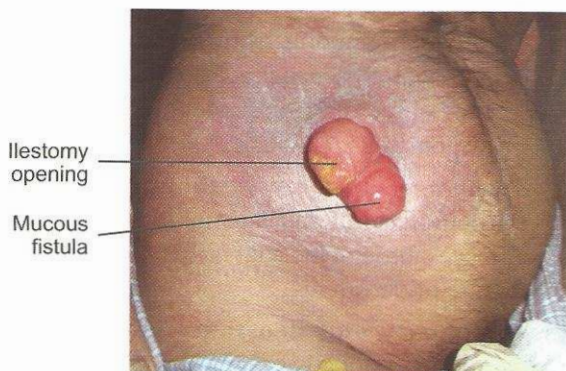


Fig. 21.14. Right iliac temporary end ileostomy with mucous fistula – note the maceration and inflammation of the skin around the ileostomy.

Methods

1. *Site of election:* Both the types are usually sited on the right iliac fossa, near the outer margin of the right rectus muscle about 5 cm. lateral to the midline and 4 cm. below the umbilicus.
2. The ileal stoma should protrude as a nipple for at least 3–5 cm above the skin surface. This facilitates the fluid effluent to pass directly into the collecting bag and thus with less chance of seeping between the wafer of the collecting bag and the peristomal skin. It prevents peristomal skin irritation and break down.
3. The edge of the ileal mesentery should be sutured to the peritoneum of lateral abdominal wall. This prevents herniation of small bowel loops through the paraileal stomal space.

4. A stoma adhesive disc is applied to the ileostomy stoma in the operation theatre. An ileostomy bag or pouch is placed over the disc. People with ileostomies either use a 'closed end' pouch which must be thrown away when full, or a 'drainable pouch' that is secured at the lower end with a leak proof clip.
5. Ostomy pouches fit close to the body surface and are usually not visible under the regular clothing unless the patient allows the pouch to become full.
6. The ileostomy starts to work within 48 hours.
7. It produces a daily output of 500 to 700 ml liquid or semi liquid contents.
8. Ordinarily the pouch must be emptied several times a day and changed every 2-5 days.

Complications of ileostomy

See complications under colostomy.

Early problems:

1. Occasional necrosis of the distal ileum due to devascularization.
2. Parastomal infection

Late problems:

1. Fistula
2. Prolapse
3. Stricture
4. Parastomal skin ulceration
5. Obstruction of ileostomy with food fibers e.g., potato skin, raw vegetables.
6. Effluent amount larger than 1000 ml/day can cause fluid and electrolyte imbalance. This requires urgent attention and treatment: correction of fluid and electrolyte imbalance, use of drugs like loperamide or lomotil, use of bulk laxatives like isopguls husk.
7. Kidney stones.
8. Gall stones.

Examination of a patient presenting with intestinal stomas

Examine:

1. The site of intestinal stoma:
 - (a) Left iliac fossa – Sigmoid colostomy
 - (b) Right hypochondrium – Transverse colostomy
 - (c) Right iliac fossa – Ileostomy
Caecostomy

2. Number of openings:
 - (a) One opening – End ostomy
 - (i) Sigmoid colostomy

$\left\{ \begin{array}{l} \text{Permanent, e.g.,} \\ \text{following APR} \\ \text{Temporary, e.g.,} \\ \text{Hartmann's} \end{array} \right.$
 - (ii) Ileostomy – permanent
 - (b) Two openings – usually temporary
 - (a) Loop ostomy
 - (i) Transverse colostomy
 - (ii) Sigmoid colostomy
 - (iii) Ileostomy
 - (b) Double-barrelled colostomy
 - (i) Devine procedure
 - (ii) Paul Mickulicz procedure
3. Protrusion or projection of the ostomy opening from the skin surface
 - (a) Colostomy – at least 2-5 cm above the skin surface
 - (b) Ileostomy – at least 3-5 cm above the skin surface
4. Colour of the stoma – usually pink red.
5. Skin condition around the ostomy opening:
 - (a) Smooth, healthy and dry appearance in comparison with the rest of the abdominal surface.
 - (b) Excoriated, macerated and wet surface with or without faecal smell.
 - (c) Irregular scar, skin folds or corrugation around the ostomy – allows seepage of the effluent onto the abdominal surface under the flange of the collecting bag which can cause maceration of the skin around the ostomy.
6. Whether the ostomy allows introduction of the patient's or examiner's index finger easily – to exclude stenosis of ostomy.
7. Examine the patient's perineum in the presence of left iliac end colostomy for linear midline scar and absence of anus. In the presence of anus do digital examination for rectum:
 - (a) Left iliac end colostomy in APR – anus will be absent.
 - (b) Left iliac end colostomy in Hartmann's procedure – anus will be present and will allow digital examination.
8. Enquire what is the consistency of the effluent:
 - (a) Colostomy – usually solid or semisolid. If history of constipation or spurious diarrhoea – exclude impaction of stool. This may require enema through colostomy or digital removal of impacted stool. Patient should be advised plenty of drinks, plenty of high fibre diet and frequent purgatives like dumphalac.
 - (b) Ileostomy – usually liquid or semi-liquid. Enquire the quantity of the daily output – usually produces a daily output of about 500–700 ml. Amounts more than 1000 ml/day cause fluid and electrolyte imbalance and increase the likelihood of stone formation.
9. Look for any complication of the ostomy.
10. Enquire for the type of collecting bags used by the patient and any problems faced by the patient. Collecting bags may be
 - Reusable colostomy bags
 - Disposable colostomy bags with or without flange
 - Ileostomy bags placed over the stoma adhesive disc.

22

CHAPTER



Orthopaedic Disorders

CONGENITAL DEFORMITIES

CONGENITAL TALIPES EQUINOVARUS

This deformity is made up of three elements:

1. *Equinus*: The heel is drawn up by tight tendo-achillis;
2. *Inversion*: The sole is pointing medially;
3. *Varus*: The forefoot is adducted relative to the hindfoot.

Aetiology

Idiopathic: Cause is unknown.

Most cases are due to a defect in the foetal development with imbalance *between the plantar-flexor-invertor and dorsiflexor-evertor groups of muscles*. Milder forms may be due to prolonged malposition of foetal foot in the uterus, but this cannot be accepted for the severe cases. An additional neuromuscular defect is possibly relevant.

Identical deformity can be seen in babies with neurological lesions such as spina bifida or poliomyelitis, or with arthrogryposis multiplex congenita where the muscles are abnormal.

Pathological anatomy

The actual defect is subluxation of the talo-navicular joint, so that the talus points downwards (*equinus*) and slightly outwards in relation to the calcaneum, while the navicular along with the entire fore foot is

displaced medially upon the head of the talus and rotated into supination (*the forefoot becomes adducted and inverted – the composite varus deformity*). There may be internal rotation of the tibia as well in severe cases.

The soft tissues (muscles and ligaments) at the medial side of the foot are also underdeveloped and contracted. In most cases calf and peroneal muscles are underdeveloped and thin as well.

In the absence of early effective treatment secondary growth changes occur in the bones perpetuating the deformity.

Diagnostic criteria

1. *Boys are more affected.*
2. Condition may be bilateral.
3. In mildest form – the forefoot alone found to be adducted relative to the hindfoot.
4. In well-formed cases – *the typical deformity consisting of three elements are noted*:
 - adduction of the forefoot;
 - inversion of the foot; and
 - plantar flexion.

The outer border of the foot is convex and the inner border is concave. The sole is directed medially. The heel is small and high. There may be deep furrow at the leg above the heel.

The skin of the foot is usually stretched over the dorsum of the foot but thrown into creases along the



Fig. 22.1. Bilateral talipes equinovarus. Note that the forefoot is adducted and inverted (the sole pointing medially). Equinus (plantar flexion) deformity is not evident from the front in this picture.

inner border and the sole (*the skin creases are absent in varieties other than in congenital one*).

The head of the talus may be seen and felt to protrude on the dorsolateral surface of the foot.

The leg may be underdeveloped and thinner than that of the healthy side. There may be some internal rotation of the tibia on its long axis and in a few cases genu valgum.

5. Passive movements of the foot are difficult and painful. (*In normal infants it is possible to evert and dorsiflex the foot far enough until the little toe touches the front of the leg.*)

6. X-rays: Shows the shape and positions of the tarsal ossific centres or bones. In AP-view of a normal foot the line projecting the long axis of the talus forwards, passes medial to the first metatarsal or coincides with it. In uncorrected club foot the line passes lateral to the first metatarsal.

In lateral view of a normal foot with full dorsiflexion the long axis of calcaneum should form an acute angle with that of the tibia and the talocalcaneal angle should be at least 20 degrees.

Treatment

Depends on the age of presentation.

Primary conservative treatment

Objectives of treatment

1. To correct the deformity early and fully by repeated manual pressure; and

2. To hold the corrected positions until the foot stops growing.

Timing

Ideally the treatment should start within one week of birth.

Correction of deformity

Each component of the deformity is corrected in turn and always in the following order:

- First, the forefoot adduction;
- Then, inversion; and
- Finally, the equinus.

It may require three to six manipulations with or without anaesthesia.

Maintenance of correction

The corrected position of the foot is held there preferably by a plaster of pairs cast after each manipulation. The plaster cast must extend to the upper thigh with the knee flexed to 90 degrees, otherwise the infant is able to draw the foot up inside the plaster cast.

There are two groups of club foot: *easy and resistant*. *Easy groups* respond readily to stretching and splinting. *Resistant groups* respond poorly, and relapse quickly even after repeated manipulations, and so these groups require early operative corrections.

Operative treatment

The resistant case is best operated upon by 6 weeks. *The operation is soft tissue release at the medial side of the foot and ankle and lengthening of the tendoachillis*. Through a medial incision all the tight ligaments and fascia at the medial side of the foot and ankle are divided and the tight tendons (the invertors) are divided. This is followed by elongation of the tendoachillis *usually by tenotomy*. Finally, the foot must be corrected fully and the corrected position held in plaster for two to three months.

Treatment after correction

Whether or not operation was needed, splintage should be continued with the Denis-Browne splint until the child starts walking; thereafter correction can be maintained by Denis-Browne splint at night and Denis-Browne shoe which permits walking in eversion during the day. Splintage may need to be continued until puberty.

Treatment in neglected or relapsed cases

Several types of operations are available to restore a plantigrade foot.

Between 2 and 5 years of age: Operations involve:

- Division of contracted soft tissues at the medial side of the foot including the medial talocalcaneal ligament and the posterior capsule of the ankle;
- Transfer of the tibialis anterior tendon to the outer side of the foot, or
- Transfer of the tibialis posterior tendon through interosseous membrane to the outer side of the foot, to supplement the action of the evertor muscles;
- Lengthening of a short tendoachillis by Z-plasty.

After the age of 5 years: Some form of bone reshaping is also required. This involves:

- Dorsolateral wedge excision of calcaneocuboid joint followed by arthrodesis to shorten the lateral border of the foot (Evans, 1961).
- Osteotomy of the calcaneum with insertion of a bone wedge in the medial side to correct the inversion of the foot (Dwyer, 1963).

Over the age of 12 years: The best operation is a dorsolateral wedge tarsectomy so that when the gap is closed the foot becomes plantigrade.

When the patient reaches puberty: A triple arthrodesis could be considered.

SOFT TISSUE LESIONS

A. DISEASES OF FASCIAE

DUPUYTREN'S CONTRACTURE

This lesion consists of fibrosis in the palmar aponeurosis that lie deep to the subcutaneous tissues of the hand and superficial to the flexor tendons, leading to flexion contracture of one or more of the fingers in late cases.

Aetiology

1. Unknown: The condition is often familial and inherited as an autosomal dominant.
2. Most common in people of European (especially Anglo-Saxon) descent.
3. The condition is often bilateral.

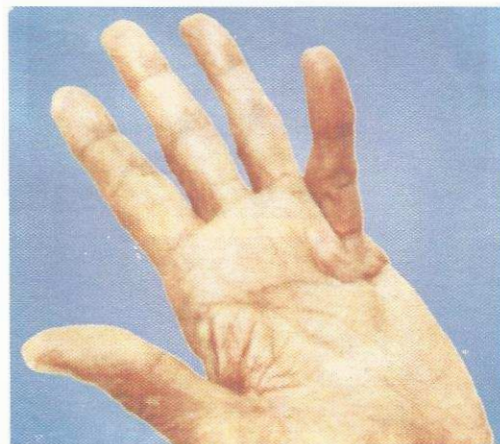


Fig. 22.2. Dupuytren's contracture—causing flexion deformity of the little finger *flexion at the MP and proximal IP joints.* (Courtesy: Dr. Yogesh Salphale, D'ortho) Note and palpate the thickened subcutaneous band from the base of the little finger to the palm.

4. In a predisposed person, repeated trauma or long continued pressure possibly plays a role but its exact significance is uncertain.
5. A higher incidence is noted among epileptics receiving phenytoin therapy. Association with alcoholic cirrhosis, diabetes and pulmonary tuberculosis has also been noted.

Surgical pathology

Due to fibrosis, the palmar aponeurosis becomes greatly thickened and gradually shrinks. In the beginning, a thickened indurated plaque or nodule appears in the palm at the level of the distal palmar crease at the base of the ring finger. The skin is also thickened and firmly attached to the fibrous nodule beneath. Very slowly the induration spreads distally in the fascia towards the ring and little finger, and proximally up the palm. *Gradually the fascia contracts, pulling the fingers into flexion at the metacarpophalangeal and proximal interphalangeal joints.* The medial half of the aponeurosis is affected most. *So the deformity is usually confined to ring and little fingers.* Occasionally middle finger may be affected. *The joints themselves are unaffected except in late cases when secondary capsular contractures occur.*

Rarely, the plantar aponeurosis in the foot is similarly affected.

Very rarely, this condition may be associated with fibrosis of the corpus cavernosum (Peyronie's disease).

Diagnostic criteria

1. Usually common in middle-aged men.
2. In early cases the patient may complain of pain on grasping, or a small nodule in the midpalm. In a late case, the patient may present with typical deformity. The condition may be bilateral.

On examination:

3. In an early case, a small thickened nodule or plaque in the palm is felt at the distal palmar crease opposite the base of the ring finger. There may be obvious subcutaneous cords extending into the ring or little finger or both, leading to flexion of the fingers at the metacarpophalangeal and proximal interphalangeal joints (Fig. 22.2). The distal interphalangeal joints remain unaffected. The cords get tighter and more prominent if the fingers are extended passively.

Eventually, there is extreme flexion of the finger with the tips digging into the palm. The flexion of the fingers is not lessened by the flexion of wrist joint (cf. Volkmann's contracture).

4. The overlying skin is closely adherent and often puckered.
5. In some cases, there may be thickening of the skin over the dorsum of the proximal interphalangeal joints. These are known as *Garrod's pad*.
6. Sole of the foot should be examined for any nodule or puckering.
7. Peyronie's disease should be considered.

Differential diagnosis

- Congenital contracture of the little finger.
- Volkmann's contracture leading to Claw hand.

Treatment

1. In early cases, this condition may be treated by gentle stretching of the fingers and night splintage.
2. In established contracture, operation is necessary.

Closed fasciotomy entails simple division of the taut contracted bands until passive correction is obtained. But there is a danger of injuring the underlying nerves and vessels; moreover the contracture tends to recur.

Open fasciectomy is the operation of choice and entails radical excision of the thickened part of the palmar aponeurosis by painstaking dissection. The affected area is approached through Z-shaped incision along the natural crease lines. Post-operatively, splintage and daily wax baths and exercises are required to restore mobility.

Amputation is occasionally advised in presence of severe contraction of the joint capsule.

CARPAL TUNNEL SYNDROME

In this condition the median nerve is compressed as it passes through the carpal tunnel — the space between the flexor retinaculum and the carpal bones.

Surgical anatomy

Carpal tunnel: The carpal tunnel is a fibro-osseous tunnel, the floor of which is formed by the concavity of the carpal bones and the roof is formed by the flexor retinaculum. Through this tunnel pass the following structures:

1. Flexor digitorum superficialis and profundus, the long flexor of the fingers with their common synovial sheath (the ulnar bursa).
2. Flexor pollicis longus, the long flexor of the thumb with its own synovial sheath (the radial bursa).
3. *Median nerve which passes deep to the retinaculum* on the lateral side of the flexor digitorum superficialis (middle finger tendon), between it and flexor carpi radialis.

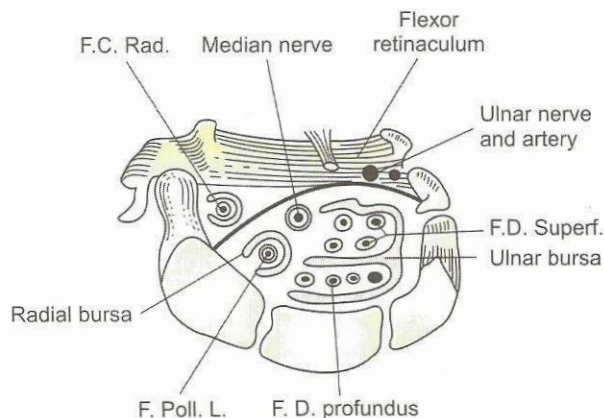


Fig. 22.3. Carpal tunnel with its contents. Note that ulnar nerve lies superficial to the retinaculum and median nerve lies deep to the retinaculum.

So, through this small, tough and non-resilient tunnel so many structures are crowded. There is normally insufficient space for the median nerve in the carpal tunnel. Any lesion diminishing the size of the compartment can cause compression of the median nerve.

Compression of the median nerve causes ischaemia of the nerve which, if prolonged, can cause ischaemic damage of the axon, resulting in paraesthesia, numbness and motor weakness in its distribution.

Since the superficial palmar branch of the median nerve is given off proximal to the retinaculum, there is usually no sensory impairment over the thenar eminence. Note also that *the ulnar nerve passes superficial to the flexor retinaculum* and so is spared.

Causes of compression

Compression can be caused by skeletal abnormalities, swelling of the soft tissues within the tunnel, or thickening of the retinaculum.

Skeletal deformities: Osteoarthritis, rheumatoid arthritis, dislocation of lunate.

Soft tissue deformities:

- Swelling of the tendon sheaths during menopause and pregnancy without any very obvious cause. Women have smaller tunnels than men.
- Rheumatoid or tuberculous tenosynovitis e.g., compound palmar ganglion.
- Lipoma in the hand.

Diagnostic criteria

1. Age and sex: *Common in middle aged women at the menopause.* In case of younger patients – pregnancy, rheumatoid disease, tenosynovitis, or skeletal deformities may be the factors.
2. The patient presents with the *complaints of pain and paraesthesia in the distribution of median nerve* i.e., in the radial three and a half digits. Little finger is never affected (as it supplied by the ulnar nerve). These symptoms are often prominent during night — *the patient is often woken in the middle of the night* with burning pain, tingling and numbness, and a feeling of stiff swollen fingers and hand. The patient may have to move the fingers or shake the hand to get relief. The pain may radiate up the inner side of the forearm.

As the lesion progresses, the patient complains of clumsiness in carrying out fine movements such as sewing, writing.

On examination:

3. The typical pains and paraesthesia can be reproduced by holding the wrist fully palmar flexed for one or two minutes or by compressing the arm with a sphygmomanometer cuff.
4. In early cases the hand looks and feels normal. In late cases there is wasting of thenar eminence with reduced sensation of the thumb, index and middle fingers.
5. Adduction and opposition of the thumb may be weak.
6. Peripheral pulses and temperature are normal.
7. Evidence of myxoedema, pregnancy, osteoarthritis, and rheumatoid arthritis should be looked for.
8. *One should exclude the other causes of paraesthesia in the hand such as* cervical spondylosis, cervical rib, peripheral neuritis.
9. *Nerve conduction studies on the median nerve at the wrist show delay in conduction and confirm the diagnosis.*
10. X-ray of the cervical spine: AP and lateral views to exclude cervical spondylosis and cervical rib.

Treatment

Operation is the treatment of choice and includes longitudinal division of the flexor retinaculum to decompress the nerve. Operation causes immediate relief of pain but neurological deficits may not be recovered fully.

B. DISEASES OF SYNOVIUM AND TENDON SHEATHS

BURSA

A bursa is a fluid-filled sac of connective tissue lined with flattened endothelium similar to synovium. It is interposed between two moving surfaces to reduce friction or is placed over prominent bony points to act as a protective cushion.

Bursa may be of three varieties:

1. True bursa
2. Bursa communicating with the joints
3. Adventitious bursa

TRUE BURSA

1. Subdeltoid bursa
2. Olecranon bursa
3. Prepatellar bursa
4. Superficial infrapatellar bursa
5. Tendoachillis bursa
6. Gluteal bursa
7. Ischial bursa

Individual bursa of clinical importance

Subdeltoid bursa

- It lies between the deltoid muscle and underlying greater tuberosity of the humerus.
- It may be affected by chronic tuberculous disease or by acute gonococcal infection.
- It forms a fluctuant tender swelling beneath the deltoid muscle and makes active abduction at shoulder painful and limited.

Olecranon bursa

- It lies between the skin and the olecranon process.
- Chronic serous bursitis is common particularly in students who are in the habit of resting their elbow over the table during their study — so known as 'Student's Elbow'.
- Also common in miners—Miner's elbow.
- It is also the seat of gouty deposits.
- The bursa may also be affected by rheumatoid arthritis, tuberculosis or rarely syphilis.

Prepatellar bursa

- Chronic serous bursitis of prepatellar bursa due to repeated minor trauma, characterised by a swelling

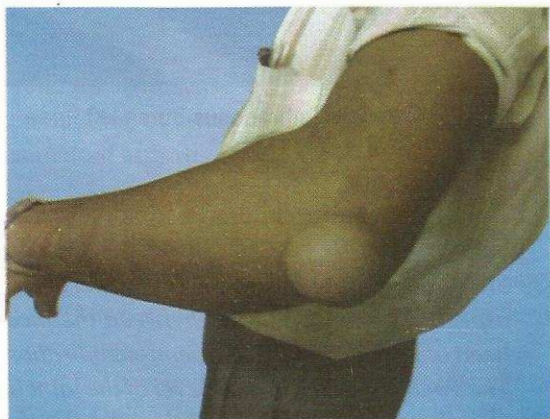


Fig. 22.4. Olecranon bursa left elbow.

in front of the patella, is common in housemaids — so known as '*Housemaid's knee*'.

- Now-a-days it is also common in miners and carpet layers. Such occupational hazards may stop the patient from working.

Superficial infrapatellar bursa

- Chronic serous bursitis of this bursa is common in clergymen who spend long hours in kneeling down during the prayer — so known as '*Clergyman's knee*'.
- It should be differentiated from Osgood-Schlatter's disease.

Gluteal bursa

- It lies between the tendon of the gluteus maximus and the greater trochanter of the femur.
- It may be rarely affected by tuberculosis.
- It forms a fluctuant swelling behind the trochanter and causes abduction and lateral rotation at the hip joint painful.

Ischial bursa

- Chronic serous bursitis of the bursa in relation to the ischial tuberosity characterised by a cystic swelling is commonly met in the weavers who sit for a long time on their ischial tuberosity — so known as '*Weaver's bottom*'.
- It can cause discomfort or pain on prolonged sitting.

BURSA COMMUNICATING WITH KNEE JOINTS

Surgical anatomy

Bursae around the knee-joint: There are about 12 bursae around the knee joint of which 4 are anterior, 2 posterior and 3 each on medial and lateral sides.

A. *Anterior bursae:* 4 in number (Fig. 22.5):

1. Suprapatellar – communicates with the knee joint.
2. Prepatellar.
3. Superficial infrapatellar.
4. Deep infrapatellar.

B. *Posterior bursae:* 2 in number (Fig. 22.6): One between each head of origin of gastrocnemius and the capsule of the knee joint. These may often communicate with the knee joint.

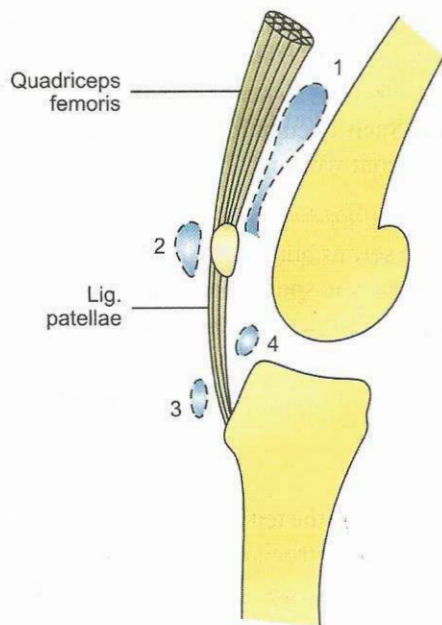


Fig. 22.5. Bursae in front of the knee joint.

C. Medial bursae: 3 in number (Fig. 22.6)

1. One lies superficial to the medial ligament of the knee – separating the sartorius, gracilis and semitendinosus from the medial ligament as these muscles cross it.
2. One deep to the medial ligament of the knee – separating the ligament from the tendon of semimembranosus.
3. One between the semimembranosus and the medial condyle of the tibia – semimembranosus bursa (5 in Fig. 22.6).

D. Lateral bursae: 3 in number (Fig. 22.6)

1. One superficial to the lateral ligament of the knee – separating the biceps tendon from the lateral ligament.
2. One between the lateral ligament and the tendon of popliteus.
3. One between the tendon of popliteus and the lateral condyle of femur – communicates with the joint.

SEMIMEMBRANOSUS BURSA

This bursa lies between the semimembranosus tendon and the medial condyle of the tibia. It very often communicates with the knee joint through a narrow aperture. Chronic serous bursitis is frequent particularly among children and adolescents.

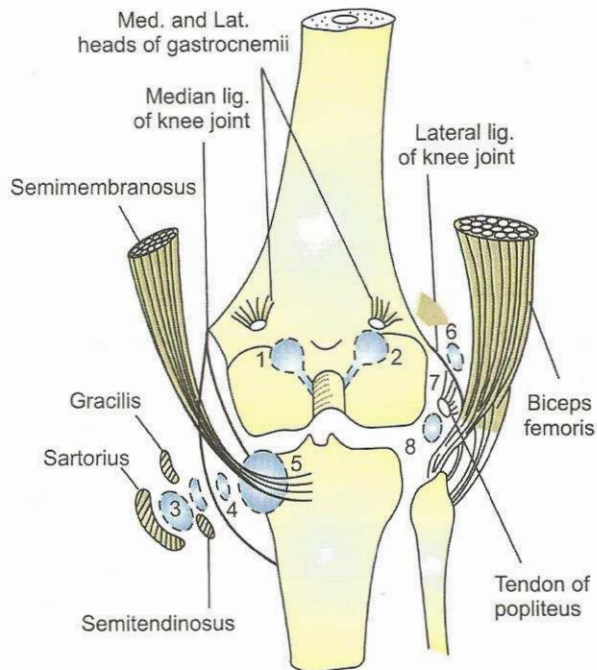


Fig. 22.6. Posterior, medial and lateral sets of bursae around the knee joint.

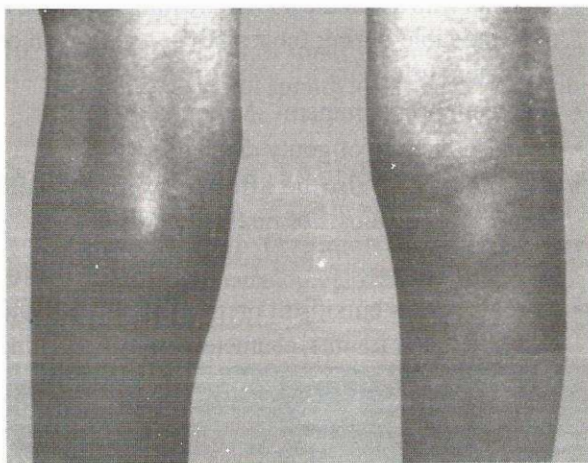


Fig. 22.7. Semimembranosus bursa left knee.

Diagnostic criteria

1. **Age:** Common in children and adolescents of both sexes.
2. **Site:** Swelling situated at the posteromedial aspect of the knee joint in between the medial head of gastrocnemius and semi-membranosus. The swelling lies above the level of the joint line, slightly to the medial side of the popliteal fossa (cf. Baker's cyst).

3. On flexion of the knee joint – the swelling becomes less prominent.
4. On extension of the knee joint (when the patient stands erect), the swelling becomes prominent. Due to contraction of the muscle, the communication of bursa with the joint cavity is closed, hence the size of the swelling becomes prominent.
5. *Cystic in feel: Fluctuation positive*, but the fluid cannot be pushed into the joint, presumably because the muscles compress and obstruct the occasional narrow aperture.
6. Transillumination – may be positive.
7. Usually no pain or tenderness.
8. Knee examination and movements are otherwise normal (cf. Baker's cyst).
9. *Non-pulsatile*: Popliteal and peripheral pulses are normal.

Differential diagnosis

1. Morrant Baker's cyst
2. Cold abscess of knee joint
3. Popliteal aneurysm – *Pulsatile swelling*
4. Lipoma
5. Sebaceous cyst.

Treatment

In children the operation may be deferred because the cyst may disappear. Nevertheless, if the swelling becomes uncomfortably large or painful it may be excised through a transverse incision in the popliteal fossa.

MORRANT BAKER'S CYST

This cyst results from herniation of the synovial membrane of the knee joint through the posterior capsule. It is not a primary condition but is always secondary to disease of the knee with persistent accumulation of synovial fluid as in osteoarthritis or rheumatoid arthritis.

Diagnostic criteria

1. Age: Near about 40 years or middle aged.
2. Patient presents with a swelling in the back of the knee with pain in the joint particularly during walking.
3. *Site: The swelling is situated near the mid-line in the popliteal fossa, below the level of the joint line and deep to the gastrocnemii muscles.* Some

of the cyst may bulge out between the heads of the gastrocnemii muscles (cf. Semimembranosus bursa).

4. *Cystic in feel – Fluctuation positive.*
5. The swelling is often reducible as the fluid can be pushed into the joint.
6. Examination of knee joint shows evidence of arthritis.
7. *Effusion in the knee joint* is obvious and can be demonstrated by patellar tap.
8. Knee movements: *Painful and restricted with or without crepitus* (cf. Semimembranosus bursa).
9. X-ray – evidences of arthritic changes in knee are present.

Diagnostic criteria

Semimembranosus bursa.

Treatment

1. Aspiration and injection of hydrocortisone followed by crepe bandage.
2. Treatment of the associated joint pathology if any e.g., rheumatoid or osteoarthritis.
3. If conservative treatment fails, excision of the cyst with or without synovectomy of the knee joint can be performed.

ADVENTITIOUS BURSAE

These are *new formations of sacs* over bony prominences secondary to repeated but prolonged friction and pressure.

Examples

1. Bursa over the medial aspect of the head of the first metatarsal bone particularly in a case of hallux valgus. This is also called *Bunion*.
2. Bursa found on the nape of the neck, opposite the spine of the 7th cervical vertebra – commonly affecting the fish-sellers or persons carrying weight on the neck. This was popularly known as *Billingsgate hump* (Billingsgate was the largest fish market in London).
3. Bursa over the shoulder – commonly affecting the persons carrying weights on the shoulder.
4. 'Basket carrier's bursa' on the scalp.
5. 'Tailor's ankle' – A bursa over the lateral malleolus.
6. Bursa developing over an exostosis.

7. Post-calcaneal bursa – develops at the back of the heel between the tuberosity of the calcaneum and the skin due to repeated friction from ill-fitting shoes.

GANGLION

A ganglion is a cystic swelling, originating in the synovial membrane of tendon sheaths or of the smaller joints due to myxomatous degeneration. The connection with the parent synovium usually cannot be demonstrated. The cyst contains a viscous jelly like material.

Common sites

1. Dorsum of the wrist – commonest.
2. Dorsum of the foot and ankle.
3. Palmar aspect of the wrist and fingers.

Cysts of the lateral semilunar cartilage of the knee are similar histologically to ganglion.

Why commonest over the dorsum of the wrist?

Due to the presence of many tendons with their sheaths.

Names of the tendons over the dorsum of the wrist:

The tendons from lateral to medial are:

1. Abductor pollicis longus } Common
2. Extensor pollicis brevis } sheath
3. Extensor carpi radialis longus } Common
4. Extensor carpi radialis brevis } sheath

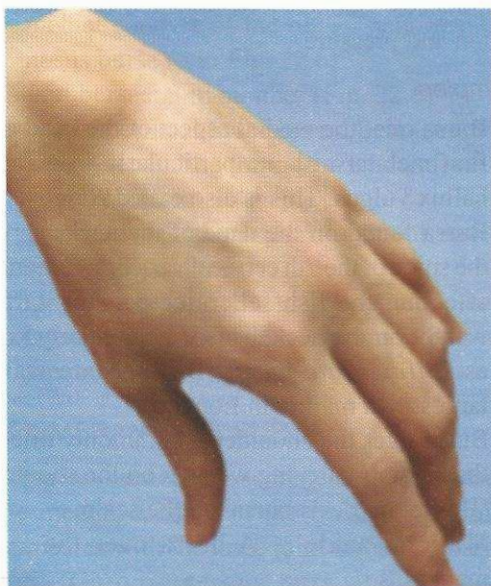


Fig. 22.8. Ganglion dorsum of the left wrist.

5. Extensor pollicis longus – Single sheath
6. Extensor indicis } Common
7. Extensor digitorum } sheath
8. Extensor digiti minimi – Single sheath
9. Extensor carpi ulnaris – Single sheath

So, there are 9 tendons within 6 sheaths.

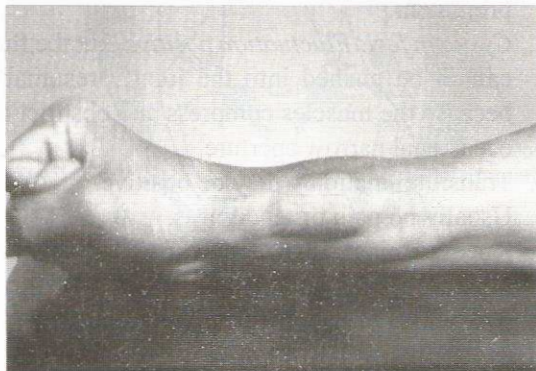


Fig. 22.9. Ganglion – involving common sheaths of the tendons of abductor pollicis longus and extensor pollicis brevis.

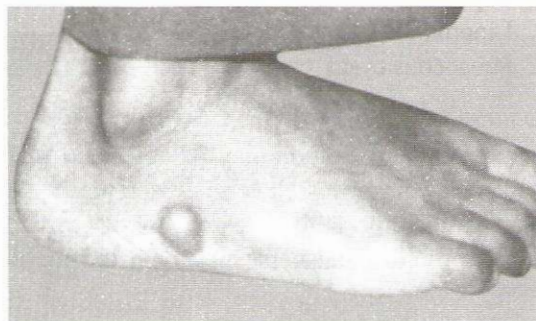


Fig. 22.10. Ganglion in relation to peroneus brevis. D/D: Implanted dermoid.

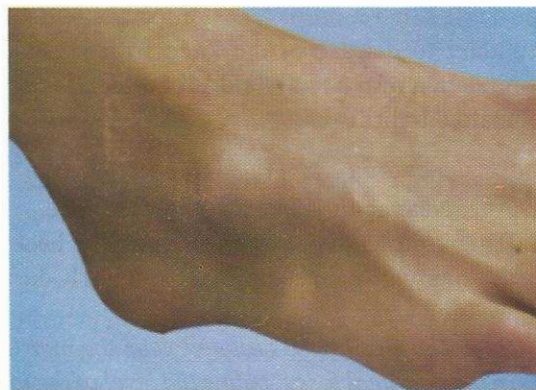


Fig. 22.11. Ganglion in relation to peroneus tertius.

Diagnostic criteria

1. Commonly found in adults. Common in females.
2. Patient presents with a lump which is usually slowly growing and has been present for months or years. Patient seeks advice either because of disfigurement, or pain particularly after strenuous work with the hand.

On examination:

3. The lump appears as a well defined spherical, firm, cystic swelling; it sometimes becomes so tense that it feels bony hard and apt to be mistaken for a tumour. Surface is smooth. Overlying skin is freely mobile over the ganglion.
4. Fluctuation: Positive (by Paget's test) in a soft swelling.
5. *Mobility*: Swelling is mobile across the axis of the tendon (i.e., on side to side movement), but not mobile along the axis of the tendon.
6. *Relation of the swelling with the tendon*: The patient is asked to make the tendon taut to find out whether the ganglion is fixed to the tendon or not. If fixed, then find out the tendon to which it is related.
7. Occasionally a ganglion may slip away between the deeper structures while pressed, giving the false impression that its contents have reduced inside the joint.

Treatment

Excision of the ganglion followed by biopsy.

Why biopsy: Frequently the synovial sheath in relation with the tendon may have neoplastic change, benign or malignant, called synovioma.

Fate of operation: It may be completely cured, or, to the annoyance of the patient, it may recur.

Other methods of treatment

1. Time-honoured treatment is rupturing the ganglion by striking it with a heavy object (*Bible treatment* used by the Priests in older days for rupturing the ganglion with the help of the Bible).
2. Aspiration of ganglion and introduction of sclerosing agents such as ethanolamine.
3. Suturing of the ganglion by multiple nylon threads.

TENDON SHEATHS

Stenosing tenovaginitis

This occurs in the fibrous sheath of the tendon and is due to either chronic inflammation or degeneration from overuse. There is thickening and constriction of the sheath with the tendon becoming thickened above and below. This is commonly found at two sites – in the wrist (De Quervain's disease) and in the finger (Trigger finger).

DE QUERVAIN'S DISEASE

Here, the common sheath covering the abductor pollicis longus and extensor pollicis brevis tendons on the lateral side of the wrist *just above the styloid process* is involved.

Diagnostic criteria

1. Commonly seen in middle aged women.
2. Patient complains of pain on the radial side of the wrist which becomes worse during wringing clothes or towels.

On examination:

3. A localised tender lump may be felt just above the radial styloid process.
4. Ulnar deviation of the wrist is painful.
5. Passive adduction of the thumb is painful.
6. Active extension and/or abduction of the thumb against resistance is also painful.
7. Crepitus may rarely be felt locally as the tendons move.

Treatment

Conservative: May be tried in early cases but recurrence is expected.

1. Rest, oral analgesics, anti-inflammatory drugs, local heat therapy and ultrasound application may hasten the progress.
2. Local injections of hydrocortisone may cure and reverse the process in early cases.

Operative: Division of the involved tendon sheath gives uniform and permanent satisfactory result.

TRIGGER FINGER

Here, stenosis occurs at the entrance to the fibrous flexor tendon sheath opposite the metacarpal heads, causing impediment to sliding of the tendon into the

sheath during extension. Several digits may be affected.

A similar condition is seen in the thumbs of infants which may be congenital (Trigger thumb).

Diagnostic criteria

1. In adults any finger may be affected – the ring and the middle fingers are commonly affected. In infants, the thumb is commonly affected.
2. Patient notices that when the hand is unclenched, the affected finger remains bent. The affected finger gets fixed in flexion until it is extended by excessive voluntary effort or made passively straight by the other hand. When extension occurs it does so with a characteristic 'snap'.
3. Locally one may find tenderness and/or a palpable nodule in front of the metacarpal head of the affected finger.

Treatment

1. Local injections of Hydrocortisone may cure and reverse the process in early cases.
2. Division of the involved fibrous sheath gives uniform and permanent satisfactory results.

Infective tenosynovitis

The synovial tendon sheaths may be infected by tubercle bacilli liberated into the blood from a primary focus elsewhere. The commonest site for tuberculous tendon sheath infection is in the common sheath of the flexor tendons of the fingers in front of the wrist (i.e., radial bursa) where the swelling extends both above and below the flexor retinaculum. Such swelling is commonly known as compound palmar ganglion.

The common sheath of the peroneal tendons at the lateral side of the ankle may also be affected.

COMPOUND PALMAR GANGLION

It is an inflammatory degeneration of the common synovial sheath that surrounds the flexor tendons of the fingers in front of the wrist and gives rise to a swelling *which extends both above and below the flexor retinaculum* – a bilocular swelling, so known as compound ganglion.

Common cause of such inflammation is tuberculosis. But it may also occur secondary to rheumatoid arthritis.

Is it a true ganglion? – No, because it is not due to mucoid or myxomatous degeneration.

Surgical pathology

Following degeneration, the common synovial sheath becomes thickened and oedematous. The endothelial lining secretes exudate which distends the sheath. There is formation of small fibrinous deposits or particles in the wall. These fibrinous particles are moulded into fine granules due to constant irritation and pressure of the moving tendons, and are cast off from the wall into the cavity. These particles look like sago-grains and are known as 'Melon seed bodies'.

When degeneration is secondary to tuberculosis, the melon seed bodies may contain tubercle bacilli, and tuberculous giant cells can be found in the wall of the synovial sheath.

Diagnostic criteria

1. Patient presents with a swelling on the anterior aspect of the lower forearm and wrist, and palm of the hand. The swelling is usually painless.
2. Patient may notice crepitus on movements of the fingers.
3. Occasionally paraesthesia due to median nerve compression may occur in the distribution of the median nerve (cf. carpal tunnel syndrome).
4. The swelling is hour-glass in shape.
5. Soft cystic or boggy in feel; surface is irregular. On deep palpation multiple nodular particles may be felt (due to the presence of melon seed bodies).
6. Fluctuation is positive.

Cross-fluctuation is also positive – Compression of the lump above the flexor retinaculum makes it distend below and vice-versa.

7. Transillumination is negative.
8. No local signs of inflammation are noted.
9. Crepitus may be felt when the patient moves his finger.
10. There may be some stiffness of the fingers and wrist.
11. Obvious wasting of the adjacent muscles may be noted due to median nerve compression.
12. There may be evidence of tuberculosis elsewhere in the body e.g., chest, other joints.

Investigations

1. Blood for Hb% TC, DC and ESR
2. Mantoux test
3. Chest X-ray: To exclude pulmonary lesion
4. X-ray of the wrist joint.

Complications

1. Paraesthesia due to median nerve compression.
2. Occasionally wasting of the adjacent muscles.
3. The tendons in relation may eventually fray and rupture.
4. Recurrence even after surgery.

Treatment

1. Anti-tubercular treatment.
2. Rest to the part by applying a plaster cast over the forearm and hand in palmar flexion.
3. If conservative treatment fails – *Surgery*: Longitudinal division of the flexor retinaculum and removal of the sheath after careful dissection is performed.

Swellings which are cross fluctuant

1. Compound palmar ganglion.
2. Plunging ranula.
3. Bilocular hydrocele (infantile hydrocele).
4. Psoas abscess pointing to femoral triangle.

C. DISEASES OF MUSCULO-TENDINOUS ATTACHMENTS

TENNIS ELBOW

This lesion is characterised by pain and tenderness at the common origin of the extensor muscles of the forearm into the lateral humeral condyle.

The cause is usually unknown. It is believed that it follows some unrecognised trauma which causes partial rupture of the aponeurotic fibres at the muscle origin, which is a region richly supplied by nerve endings. The elbow joint itself is unaffected. Some believe such lesion may be due to rheumatoid arthritis. Rarely the onset follows a strain during serving at tennis game.

Diagnostic criteria

1. The patient complains of *pain at the lateral aspect of the elbow* particularly on certain movements such as pouring out tea, turning a tight door-handle, or lifting with the forearm

pronated e.g., lifting articles from the roof of an almirah or carrying a bucket full of water.

On examination:

2. *Tenderness precisely localised to the front of the lateral epicondyle of the humerus.*
3. Pain is aggravated by passive stretching of the extensor muscles e.g., when the wrist is palmarflexed after holding the elbow straight with the forearm pronated.
4. Pain is also aggravated on active contraction of the extensor muscles e.g., the patient is asked to dorsiflex his wrist against resistance after holding the elbow straight with forearm pronated.
5. Movements of the elbow are full.
6. X-rays do not show any bony changes.

Treatment

1. If left alone, the symptoms may subside spontaneously. Oral analgesics, local heat therapy and ultrasound application may hasten the progress.
2. Local injection of mixture of hydrocortisone, 1 percent lignocaine, and hyaluronidase into the tender spot may give relief.
3. Operation may be necessary for persistent or recurrent cases. The origin of the common extensor muscle is stripped from the lateral epicondyle.

GOLFER'S ELBOW

This is analogous to tennis elbow except that *the common flexor origin into the medial epicondyle is affected.*

Treatment is similar.

PLANTAR FASCITIS

This lesion affects the attachment of the plantar aponeurosis to the medial tubercle of the calcaneum and causes pain and tenderness beneath the heel. The lesion is believed to be inflammatory either from repeated trauma or from rheumatism.

Diagnostic criteria

1. Common in middle aged men.
2. The patient complains of pain beneath the heel on standing or walking. The pain is more in the early morning on first weight bearing after waking up on the bed (starting trouble).

3. *On palpation* – there is marked tenderness over the site of attachment of the aponeurosis to the calcaneum.
4. Movements of the ankle are full and painless.
5. X-ray of the calcaneum: Lateral view may show a sharp spur projecting forwards from the tuberosity of the calcaneum.

Treatment

Conservative

1. Use of soft hawai chappal, or shoe or chappal with excavated heel filled with sorbo-rubber.
2. Local injections of hydrocortisone into the tender area may be successful.

Operative

Rarely required. The planter aponeurosis is stripped from the medial tubercle of the calcaneum.

D. DISEASES OF MUSCLES

MYOSITIS OSSIFICANS TRAUMATICA

This lesion consists of *heterotopic bone formation in the fleshy part of the muscle* causing restricted movements of the nearby joint.

Causes

The cause is unknown but it is thought to be related to muscular damage either following severe direct injury, or too early and too vigorous joint movements, particularly passive movements.

Site

Commonest site is the elbow: the muscle affected is the brachialis muscle following an injury such as supracondylar fracture of the humerus or dislocation of the elbow.

The other muscle affected is the quadriceps femoris following fracture of the femur or after a kick on the front of the thigh.

Surgical pathology

Basic pathology is ossification in a haematoma in the soft tissues near a joint. Following injury, there is avulsion of the periosteal attachment of some muscle, tendon or ligament resulting in haematoma. Subsequently the periosteal cells proliferate and liberate osteoblasts into the haematoma and ossify the haematoma.

Diagnostic criteria

1. Common in children and adolescents.
2. The patient or parents usually recalls the nature of previous injury and can relate the loss of joint movements to the injury.
3. The patient complains of stiffness of joint, with pain on forced movements.

On examination:

4. *Site*: Common sites are the lower part of the brachialis muscle at the elbow, or the lower part of the quadriceps femoris muscle at the knee.
5. *Elbow*: The elbow is fixed in flexion. Extension is restricted. Passive extension is restricted by pain. If the intramuscular ossification is extensive the joint may become completely fixed.

Knee: The knee is fixed in extension. Flexion is restricted. Passive flexion is restricted by pain.

6. *On palpation*: A bony hard mass is felt which is fixed to the underlying bone. The mass is often mistaken for a bony swelling. The surface is smooth but irregular with a normal local temperature. The muscle fibres can be felt running into the mass.
7. X-ray of the elbow: Will show a fluffy mass of calcification in front of the joint in the early stage. In late cases mature bone becomes evident.

Treatment

Complete immobilisation in plaster of paris cast for many weeks until the greater part of the callus is reabsorbed and pain disappears. The mass becomes smaller and more discrete. Afterwards the plaster is removed and gradual active non-weight bearing movements are advised. *No passive movements should be allowed.*

If elbow movements are still seriously restricted, the smaller and discrete mass can be removed.

Other types of myositis ossificans

Two other types are encountered clinically:

1. Myositis ossificans progressiva
2. Myositis ossificans circumscripta.

Myositis ossificans progressiva

This is a rare congenital lesion characterised by ectopic bone formation in the soft tissues, chiefly in the head, neck and trunk, with consequent limitation of movement. It is often associated with undue

shortness of the big toes and sometimes also of the thumbs (microdactyly).

The ectopic bone is formed by metaplasia of the connective tissue, but not from the displaced osteoblasts.

The disease usually appears in early childhood with the appearance of fever and soft tender swellings in the neck and trunk. The soft swellings gradually turn into hard masses of bone which lie in the course of the muscles, ligaments, and fascia. The ossification gradually extends, limiting movements of the spine and ribs more and more until in the worst cases there may be total immobility. Death from asphyxia may follow the immobilisation of the respiratory muscles.

No curative treatment is yet available. Systemic treatment with diphosphonates along with excision of bony bars may retard or delay the progress, but complete cure is impossible.

Myositis ossificans circumscripta

This condition is often seen in patients with brain or spinal cord damage. Heterotopic ossification is seen at several joints especially the hips, knees and shoulders leading to limitation of movements, sometimes totally stiffening the joints.

VOLKMANN'S ISCHAEMIC CONTRACTURE

This lesion consists of fibrosis of the muscles secondary to ischaemia of the striated muscle and nerve tissue (structures most sensitive to anoxia) resulting in contracture of the long flexor muscles of the forearm. This contracture leads to flexion deformity of the wrist and fingers.

Causes

Common causes of ischaemia are:

1. Direct damage to the brachial artery near the elbow at the time of injury (e.g., following supracondylar fracture).
2. Tight forearm bandage or plaster—compartment syndrome.

Both lead to tense oedema of the soft tissues of the forearm constrained within an unyielding fascial compartment.

Surgical pathology

The tissue oedema within the fascial compartment causes ischaemia to the striated muscles and nerves, and, if continued, leads to *ischaemic necrosis of the muscles and subsequently fibrosis and contracture*

of the muscles. The muscles involved are mainly flexor digitorum profundus and flexor pollicis longus. There is also temporary, or rarely permanent *ischaemic paralysis of the peripheral nerves mainly median nerve.* The nerve may be capable of regeneration but *the muscle once infarcted can never recover — it is replaced by fibrous tissue which contracts* — the essential pathology of Volkmann's contracture.

Similar ischaemia may also affect the calf muscles following injury to the popliteal artery, or tight leg plaster.

Occasionally ischaemia may affect the intrinsic muscles of the hand and foot.

Diagnostic criteria

1. Age: Usually common in children & young adults.
2. The patient presents with a fully developed deformity called 'Claw hand'.
3. The patient or parents usually recalls the cause of the deformity and can clearly relate the loss of finger extension to previous injury.

On examination:

4. All the fingers are flexed at the proximal and distal interphalangeal joints – a 'claw hand' deformity. The forearm looks thin and wasted. The hand looks pale and wasted.
5. The forearm muscles feel hard and taut. The hand feels cooler.
6. The patient is unable to extend the wrist and fingers together.
7. When the wrist is fully palmarflexed, the fingers can be straightened. When the wrist is moved into full dorsiflexion, the fingers become progressively more flexed.
8. There may be limitation of supination of the forearm due to contracture of the pronator teres.
9. *Sensory loss is usually absent.*

Differential diagnosis

1. Combined ulnar and median nerve palsy: *Sensory loss is usually evident.*
2. Dupuytren's contracture.

Treatment

Prophylactic treatment in the incipient stage is best. (But it is not described here.)

Established cases: Restoration to normal is hardly possible.

In mild cases: Acceptable function may be expected by intensive exercises guided by a physiotherapist, and graduated movements on an adjustable splint.

(The splint has a hinge at the level of the wrist and a ratchet for varying the angle. The forearm, hands and fingers are fixed in the splint in a position of full flexion of the wrist, thus allowing the fingers to straighten. The angle of flexion at the wrist is decreased daily by the screw, the fingers being firmly fixed in extension. In this way the muscles are slowly stretched and finally the fingers can be kept extended even when the wrist is dorsiflexed).

In advanced cases: Operation is essential. Two types of operations are available at present:

- A. Stripping of the common origin of the flexor muscles from the medial epicondyle and the upper ends of the radius and ulna followed by hyperextension of the hand and the fingers – *this allows the muscle origin to move distally*. The muscles obtain, in time, a new origin lower down the forearm – this is known as *Max Page Operation*. The deformity is reduced but function is not much improved.
- B. Excision of the necrotic muscles with subsequent transfer of a healthy muscle (for example, a wrist extensor or flexor) to the tendons of flexor digitorum profundus and flexor pollicis longus, i.e., *muscle transfers* – this helps in considerable improvement in function as well as correction of deformity. The muscle transfer may be supplemented with arthrodesis of the wrist joint.

When there is irreparable damage of the median nerve in extreme cases, nerve grafting could be considered as well for better improvement.

E. DISEASES OF TENDON

RUPTURED BICEPS TENDON

This is a pathological tear of the tendon of the long head of biceps at the bicipital groove of the humerus. Such tear occurs due to weakness of the tendon following repeated friction against osteophytes in an osteoarthritic shoulder. Some orthopaedic surgeons consider it a simple tear through an area of avascular degeneration. *In either case tear occurs without any violent stress or injury* (cf. supraspinatous tendon and

extensor pollicis longus tendon where also rupture occurs without any violent strain).

Diagnostic features

1. The patient is usually middle aged.
2. Gives history of sudden pain and 'snap' at the shoulder while lifting or pulling with the same arm. Soon the pain disappears and good function of the shoulder returns. Later, the patient may notice an unusual bulge of the muscle in front of the arm.

On examination:

3. Soon after the rupture there is slight tenderness at the bicipital groove. Later, when the patient is asked to flex his elbow against resistance the affected biceps belly is seen to contract and bunch up into a round lump like a ball leaving a gap proximal to it. The shoulder and elbow movements are usually otherwise normal.

Treatment

As the disability is minimal, no treatment is required. If the patient demands, operation can be performed – the distal stump of the tendon is sutured to the walls of the bicipital groove; the proximal stump is ignored.

RUPTURED TENDOACHILLIS

This is a pathological tear of the tendoachillis 5 cm above the insertion of the tendon. Such tear occurs through an area of avascular degeneration. The rupture is nearly always complete.

Diagnostic features

1. The patient is usually middle aged.
2. The patient gives the history of sudden agonising pain at the back of heel while running or jumping. He may believe that he has been struck just above the heel. He is able to walk, but with a limp.

On examination:

3. There is tenderness at the site of rupture.
4. A gap can be seen and felt in the course of the tendon 5 cm above its insertion. This gap is more prominent with dorsiflexion of the ankle.
5. *The patient is unable to tiptoe on the affected leg* – the patient is asked to raise the heel from the ground while standing upon the affected leg only. The patient is unable to do this when the tendon is ruptured.

6. Plantar-flexion of the foot is weak, and is not accompanied by tautening of the tendon.
7. **Simmond's test:** When doubt exists, this test is helpful. With the patient lying in prone position, the calves are squeezed — on normal side the foot is seen to plantar flex, but on the ruptured tendon side the foot remains still.
8. Ankle movements are normal.

Treatment

Non-operative treatment is preferred particularly in sedentary or elderly patients. Plaster immobilisation for 8 weeks with the foot in equinus is required. This is followed by wearing of shoe with raised heel for a further 6 weeks.

Operative treatment may be advised in fresh rupture particularly in athletes, to reduce the risk of lengthening of the tendon and consequent loss of "spring off". Operation entails repair of the tendon, preferably by sutures of stainless steel wire. This is followed by plaster immobilisation with the knee in flexion at 90 degrees and the foot is equinus for 6 weeks.

If the rupture is neglected for 4 weeks, conservative treatment by graduated exercises for the calf muscles is to be preferred.

MALLET FINGER

This is a fixed flexion deformity of the distal interphalangeal joint of a finger. This results from either a rupture of extensor tendon at its insertion or an avulsion fracture of the base of the distal phalanx. It occurs if the finger tip is forcibly bent during active extension — for instance, during making a bed, or by a blow on the tip of a finger by a ball (hence also known as 'baseball' finger).

Diagnostic features

1. The patient usually remembers the original injury.
2. The patient is unable to extend the distal interphalangeal joint fully.
3. The deformity is disfiguring.

On examination:

4. When the patient holds out his hand, with the fingers extended, the peculiar deformity is obvious — the distal phalanx of the affected finger remains 15°-20° flexed.

5. The surgeon can passively extend the distal phalanx, but the patient cannot extend it actively.
6. X-ray of the affected finger is required to see the avulsion fracture of the base of the distal phalanx.

Treatment

Only helps at or just after the injury.

If X-ray reveals no fracture: the finger is immobilised for 6 weeks in a splint or plaster with the distal interphalangeal joint hyperextended, and the proximal interphalangeal joint flexed at 80 degrees. The splint used is either Oatley splint or Mallet finger splint.

If X-ray reveals an avulsion fracture, operative fixation may be required particularly in players — the fragment is sutured back, or fixed back with a thin piece of Kirschner wire.

Once the injury is more than 3 weeks old — operative fixation rarely helps. So, the deformity can be ignored as it is minimal and functional loss is also minimal.

BONY LESIONS

CHONDROMA

This is a benign tumour composed of cartilage. It arises from the precartilaginous cells of the bone which fails to become ossified.

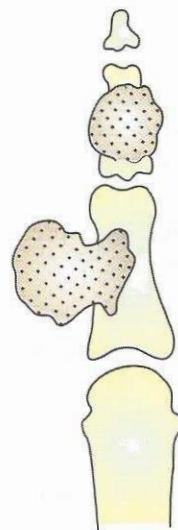


Fig. 22.12. Chondroma — Enchondroma on middle phalanx; Echondroma on proximal phalanx.

Chondroma usually involves the short long bones e.g., metacarpals, metatarsals and proximal phalanx. It can also affect the large long bones and flat bones.

Chondroma is commonly solitary.

When the chondromata are multiple and congenital (but not familial) the condition is known as *dyschondroplasia* (multiple chondromatosis or Ollier's disease). This condition begins in childhood and the large long bones are also affected where the involvement of the growth plate will interfere with normal growth of the bones leading to shortening or deformity.

Rarely chondroma undergoes malignant change and this occurs usually in one of the large long bones.

Chondroma are of two types:

1. Enchondroma
2. Ecchondroma

Enchondroma

Here the tumour grows within the bone and as it increases in size, it causes expansion and thinning of the cortex, thus forming a fusiform swelling. Fracture is common because of thinning of the cortex.

Ecchondroma

Here the tumour grows outwards on the surface of the bone.

Diagnostic criteria

1. Age: Usually present in adolescents.
2. The patient usually presents with a gradually expanding painless swelling of the bone (enchondroma), or a lump appearing on the side of the bone (ecchondroma). An ecchondroma may interfere with joint and tendon movement. The patient often may present with a fracture after a trivial injury.

On examination:

3. *Site*: Common in the short long bones of hand and feet.

If dyschondroplasia the large long bones may also be affected.

4. *Shape*: Enchondroma causes a fusiform swelling of the shaft of the bone. Ecchondroma forms a sessile lump on the surface of the bone.

5. *Surface*: Smooth. The overlying skin has a normal temperature.
6. *Consistence*: Usually hard because a chondroma is covered by a thin layer of cortical bone. The swelling is non-tender.
7. X-ray shows a well-defined, expanded rarefied area with characteristic specks of calcification.

Differential diagnosis

1. Simple cyst: There will be no specks of calcification in the rarefied area noted in X-ray.
2. Tuberculous dactylitis: There will be presence of tenderness on the swelling with inflammatory involvement of the skin. X-ray does not show specks of calcification.

Treatment

The tumour with its lining capsule should be excised and the cavity is filled up with bone chips.

Fracture, when it occurs, usually unites uneventfully and is often followed by spontaneous cure.

OSTEOMA

The word osteoma is applied to any bony overgrowth, whether or not this is a tumour. Two types are encountered clinically:

1. Compact osteoma
2. Osteoid osteoma



Fig. 22.13. Compact osteoma arising from the frontal bone. It is sessile hard lump, fixed to the underlying bone. The lump is not tender. Overlying skin is free from the lump.

Compact osteoma

Also known as *ivory osteoma*. It is a benign tumour composed of hard compact bone. *It arises in the bones which are ossified from membrane, e.g., skull bones.* It forms a smooth rounded swelling on either the inner or the outer surface of the skull bones.

When it grows on the inner surface, it causes pressure upon the cerebrum giving rise to symptoms, e.g., focal epilepsy. It is also known to develop in the orbital cavity or one of the paranasal sinuses.

When it grows on the outer surface, it appears as a visible painless lump.

Diagnostic criteria

1. The patient presents with a painless lump on the surface of the vault of the skull, frequently the forehead. The lump is very slowly growing and may have been present for many years. The lump causes disfigurement or anxiety.
2. The lump is usually not more than 1 cm.
3. The lump is sessile, flattened mounds with smooth surface.
4. *Very hard*: Hence the name ivory osteoma. It is *not tender*.
5. *Fixed to the underlying bone*, of which it is an integral part. *The overlying skin is free from the lump.*
6. X-ray: shows a sessile plaque of very dense bone with a well circumscribed margin.

Treatment

It may either be left alone or excised according to the symptoms in each case. A small area of surrounding healthy bone has to be excised with the tumour as the tumour is too hard.

Osteoid osteoma

It is a benign tumour composed of osteoid tissue with trabeculae of newly formed bone, in a vascular connective tissue groundwork. *It can arise in any bone except the skull bone.* It is common in the femur or tibia and appears as a small (usually less than 1 cm in size), round or oval lesion encased in dense bone.

Diagnostic criteria

1. The lesion is common in young males.
2. Common in the lower limbs.

3. *The leading symptom is local pain* with or without visible swelling. The pain may be severe and not relieved by rest. The patient may limp.
4. X-ray shows a small round or oval radiolucent area with sclerosis at the margin.

Differential diagnosis

The tumour is sometimes difficult to differentiate from Brodie's abscess, Ewing's tumour and chronic periostitis, and the doubt is resolved only after biopsy.

Treatment

Excision of the affected area cures the condition with relief of pain.

EXOSTOSIS (Syn. Osteochondroma)

This, the commonest benign tumour of the bone, is truly a hamartoma. It consists of normal bone covered by a cap of cartilage. Some considered it as cancellous osteoma.

Origin

It arises in childhood from a rest of growing epiphyseal cartilage plate and continues to grow on the surface at the end of the long bone *until the parent epiphysis fuses*. As the bone grows in length the tumour gets 'left behind' and thus appears to migrate along the shaft towards its centre. *When it grows, it points away from the growing end of the long bone.*

Surgical pathology

It arises at the metaphyseal regions of the long bones. It may arise from the flat bones e.g., the pelvis or the scapula.

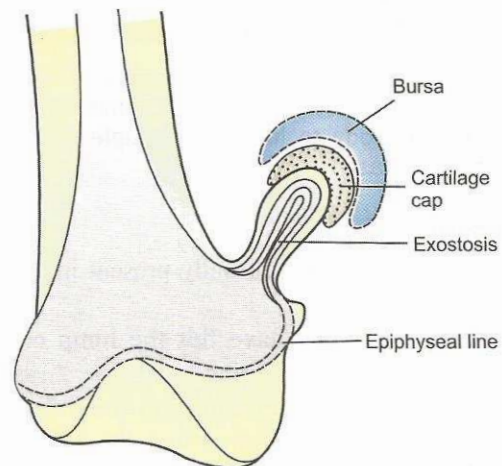


Fig. 22.14. Site of the origin of exostosis.

Exostosis consists of normal bone covered by a cap of cartilage. *It itself does not undergo malignant change*, but, rarely the cartilage cap continues to grow in adult life and gives rise to chondrosarcoma.

Occasionally adventitious bursa may develop over the cartilage cap.

Osteochondroma is usually solitary.

When they are multiple, congenital and familial, the condition is known as diaphyseal aclasis (multiple hereditary exostoses). In severe cases of diaphyseal aclasis there is interference with the skeletal growth and the patient may be deformed or dwarfed.

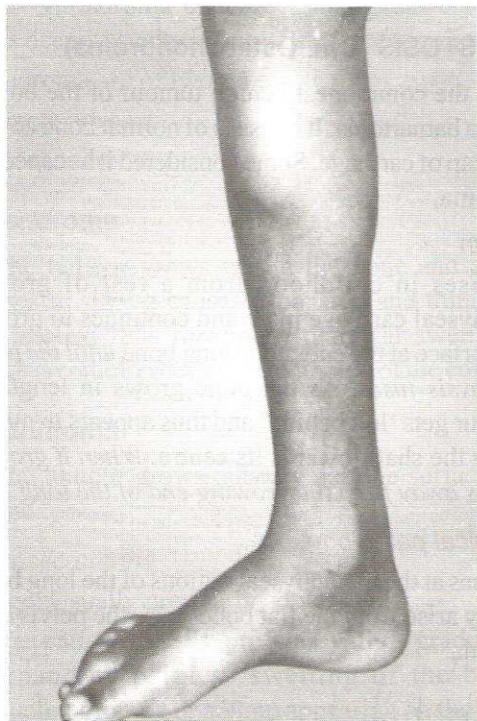


Fig. 22.15. Stony hard swelling upper end of right tibia. Exostosis (Cancellous osteoma). Examine the other joints of the body to look for multiple exostoses (diaphyseal aclasis).

Diagnostic criteria

1. Age: The patient is usually present in teenage and adult life.
2. The patient may have felt the lump or may present with a disfiguring lump.
The patient may present with the symptoms arising due to interference with the movements

of the joint and its tendons. *He may feel 'clicks' as the tendons slip over the lump.*

The lump is usually painless. But the patient may complain of neuralgia due to pressure on the adjacent nerves by the lump.

3. *Site:* The majority occur at the lower end of the femur & upper end of the tibia, i.e., around the knee.
4. *Shape and size:* The lump is pedunculated or sessile with smooth, mushroom-shaped upper end, usually 1-2 cm in diameter.
5. *Consistence:* Bony hard and non-tender. *Rarely the exostosis is covered by a soft fluctuant adventitious bursa over the cap.*
6. *Mobility:* The lump is fixed to the bone but not muscle or skin.
7. While palpating the lump, one can feel the slipping of the overlying tendon on moving the joint.
8. The rest of the bone and the nearby joint should be normal.
9. *Examine the other joints of the body* e.g., shoulder, wrist, finger to look for multiple exostoses (diaphyseal aclasis).
10. X-ray shows the mushroom like bony tumour but it looks smaller than it feels because the cartilaginous cap is invisible.

Complications

1. Fracture at the pedicle.
2. Symptoms due to pressure on the nerves (paraesthesia) or on the vessels.
3. Interference with tendon or joint movements.
4. Cartilaginous cap may become malignant – Chondrosarcoma. *Appearance of pain in an exostosis in an adult suggests malignancy.*
5. Adventitious bursa developing over the cartilage. It may be inflamed giving rise to pain and local inflammation.

Treatment

When necessary, either because of pain, or symptoms due to pressure on the nearby structures, the tumour should be excised. But one should wait until the cessation of skeletal growth (i.e., after epiphyseal fusion).

N.B. Malignant bone tumours are not discussed in this chapter because they are not given in the clinical examination. So the readers are advised to consult any textbook surgery or orthopaedics for this.

A

- Acrocyanosis, 238
- Actinomycosis, 140
- Cervicofacial, 141
- Adamantinoma, 134, 137
- Adenolymphoma, 121
- Adiposis dolorosa, 275
- Adson's test, 242
- Allantois, 158
- Ameloblastoma, 134
- Aneurysm, 244
 - Arteriovenous, 251
 - Dissecting, 245
 - False, 245, 250
 - Fusiform, 245
 - Saccular, 245
 - True, 244
 - Varicose, 251
- Aneurysmorrhaphy, 250
- Aneurysmal varix, 251
- Arteriovenous fistula
 - Aneurysm, 251
 - Iatrogenic, 253
- Branchial cartilage, 90
- Branchial cyst, 86
- Branchial fistula, 88
- Branchial sinus, 88
- Branchogenic carcinoma, 90
- Branham's sign, 252
- Bubonocoele, 167
- Bursa, 324
 - Adventitious, 327
 - Gluteal, 325
 - Ischial, 325
 - Olecranon, 325
 - Prepatellar, 325
 - Semimembranosus, 326
 - Subdeltoid, 325
 - Superficial infrapatellar, 325
 - True, 325
- Burkitt's lymphoma, 114
- Buschke-Lowenstein tumour, 227

C

- Cafe-au-lait patch, 73
- Cafe-au-lait spots, 41
- Callosities, 17
- Carcinoma
 - Cheek, 155
 - Lips, 153
 - Penis, 221
 - Tongue, 145
- Carotid body tumour, 95
- Carpal tunnel syndrome, 323

- Cattle's operation, 187
- Cavernous haemangioma, 33
- Cell nests, 55
- Cervical auricle, 90
- Cervical lymphadenopathy, 104
- Cervical rib syndrome, 239
- Cervicodorsal sympathectomy, 237
- Cheek carcinoma, 155
- Chemodectoma, 95
- Chemoreceptor, 95
- Chilblains (perniosis), 266
- Chondroma, 335
- Chordee, 294
- Cleft lip, 286
- Cleft palate, 287
- Clergyman's knee, 325
- Cockett and Dodd operation, 263
- Cock's peculiar tumour, 22
- Cold abscess in the neck, 98
- Collar-stud abscess, 101
- Condyloma acuminatum, 226
- Corn, 17
- Costoclavicular syndrome, 243
- Cryopathic ulcer, 266
- Cryptorchidism, 197
- Cyst of the epididymis, 207
- Cystic hygroma, 90

D

- Dental cyst, 133
- Dentigerous cyst, 134

Dercum's disease (Adiposis dolorosa), 25

Dermoid

- Cervical, 79
- Cyst, 18
- Implantation, 21
- Sequestration, 18
- Sublingual, 79
- Terato, 21
- Tubulo, 21

Desmoid tumour, 161

Dequervain's disease, 329

Dumbbell-shaped tumour, 42

Dupuytren's contracture, 322

E

Ectopia vesicae (Exstrophy of the bladder), 298

Ectopic salivary tumour, 129

Ectopic testis, 199

Elephantiasis scrotum, 231

Encysted hydrocele of the cord, 210

Enterocystoma, 159

Epigastric hernia, 193

Epigastric lipoma, 193

Epispadias, 297

Epithelioma, 53

Epulis, 131

Carcinomatous, 133

Fibrous, 132

Granulomatous, 132

Myeloid, 132

Erythrocyanosis frigida, 275

Erythroplasia of Queyrat, 227

Exomphalos, 189

Major, 189

Minor, 189

Exostosis, 337

F

Fatty hernia of the linea alba, 193

Fegan's test, 259

Fibrosarcoma, 161

Filariasis, 229

Fournier's gangrene, 216

Freckle (lentigo maligna), 61

Frey's syndrome, 126

Frostbite, 266

Froment's sign, 241

G

Ganglion, 328

Compound palmar, 330

Ganglioneuroblastoma, 37

Ganglioneuromas, 37

Gastroschisis, 190

Giant-celled reparative granuloma, 136, 137

Glomus body, 36

Glomus tumour (glomangioma), 36

Golfer's elbow, 331

H

Haemangioma, 31

Arterial (plexiform), 34

Capillary, 31

Venous (cavernous), 35

Hamartoma, 30

Hernia, 163

Bubonocele, 167

Congenital, 179

Epigastric, 193

Femoral, 180

Funicular, 167

Incisional, 183

Inguinal, 166

Direct, 168

Indirect, 167

Interstitial, 178

Littre's, 178

Lumbar, 194

Maydl's, 178

Obturator, 195

Ogilvie, 178

Postoperative, 183

Prevesical, 178

Richter's, 178

Sliding, 177

Spigelian, 194

Vaginal, 168

Ventral, 183

Umbilical, 188

Adult, 191

Congenital, 190

Infantile, 190

Herniotomy, 174

Herniorrhaphy, 174

Hernioplasty, 175

Hodgkin's disease, 96, 106, 109

Horner's syndrome, 238

Hourglass dermoid, 20

Housemaid's knee, 325

Hydrocele, 208

Congenital, 208

Encysted hydrocele of the cord, 210

Filarial, 212

Funicular, 209

Infantile, 209

Vaginal, 210

Hypospadias, 293

I

Intestinal stomas, 309

Colostomy, 309

Temporary

Loop colostomy, 311

Double-barrelled colostomy, 312

Devine, 312

Paul-Mickulicz, 313

Permanent

End colostomy, 313

Caecostomy, 315

Ileostomy, 317

K

Keel operation, 187 ??

Keloid, 45

Killian's dehiscence, 93

Keetley-Torek technique, 203

Klippel Trenaunay-Weber syndrome, 31

L

Ladd & Gross technique, 203

Laryngocele, 94

Leukoplakia, 144

of tongue, 144

of glans penis, 227

Lipoma, 21

Epigastric, 193

Extradural, 30

Fibro, 25

Intermuscular, 26

Intraglandular, 30

Naevo, 25

Neuro, 25

Pre-peritoneal, 193

Subcutaneous, 25

Subfascial, 26
 Submucous, 26
 Subserous, 26
 Subsynovial, 26
 Lips
 Carcinoma, 153
 Cleft, 286
 Lymphangioma, 37
 Lymphoedema, 274
 Lymphoma, 108
 Burkitt's, 114
 Hodgkin's, 109
 Non-Hodgkin's, 114
 Lymphosarcoma, 107

M

Macroglossia, 146
 Madura foot, 141
 Mallet finger, 335
 Marjolin's ulcer, 56, 266
 Median thyroid diverticulum, 80
 Melanocytic tumour, 56
 Melanoma, 56
 Aeral lentiginous, 63
 Amelanotic, 67
 Benign, 58
 Cutaneous, 61
 Juvenile, 59
 Lentigo maligna, 63
 Malignant, 61
 Melanoma in situ, 65
 Mucosal, 72
 Nodular, 63
 Ocular, 73
 Plantar, 63
 Subungual, 63
 Superficial spreading, 63
 Meningocele, 281
 Meningomyelocele, 282
 Mickulicz disease, 130
 Mixed parotid tumour, 122
 Marrant Baker's cyst, 327
 Mucous cyst, 143
 Myelocele, 284
 Myositis ossificans, 332

N

Naevus
 Blue, 60
 Compound, 59

Congenital dysplastic, 60
 Flammeus (Portwine stain), 31
 Intradermal, 59
 Junctional, 58
 Spitz, 59
 Nasopharyngeal isthmus, 290
 Neuroblastoma, 37
 Neurofibroma, solitary, 38
 Neurofibromatosis
 Cutaneous (Molluscum
 fibrosum), 42
 Elephantiasis, 41
 Generalised (Von
 Recklinghausen's disease),
 39
 Plexiform (Patchy dermatocele),
 41
 Neuroma, 37
 Acoustic, 42
 Amputation, 42

O

Odontomes, 133
 Ombradanne technique, 203
 Omentocele, 172
 Omphalocele, 188
 Orchidopexy, 202
 Osteoclastoma jaw, 137
 Osler Rendu-Weber syndrome, 31
 Osteoma, 336
 Osteomyelitis jaw (Necrosis of
 jaw), 137

P

Palomo operation, 216
 Pancoast syndrome, 243
 Papilloma, 17
 Para-umbilical hernia, 191
 Patey's faciovenous plane, 118
 Patey's superficial parotidectomy,
 124
 Operation, 109
 Parotid carcinoma, 124
 Parotid fistula, 125
 Parotid gland, 117
 Parotid swelling, 119
 Parotid tumour, 120
 Pectoralis minor syndrome, 243
 Perthes test, 260
 Peutz-Jeghers syndrome, 73

Pharyngeal pouch, 93
 Pharyngoplasty, 293
 Phimosis, 219
 Pilonidal fistula, 48
 Pilonidal sinus, 48
 Plantar fasciitis, 331
 Pleomorphic adenoma, 122
 Post-burn contracture, 43
 Potato tumour, 96
 Pratt's test, 259
 Pre-peritoneal lipoma, 193
 Pulsion's diverticulum, 93
 Pyogenic granuloma, 74

R

Ranula, 77
 Oral, 78
 Plunging, 78
 Raspberry tumour, 160
 Raynaud's disease, 235
 Raynaud's phenomenon, 234
 Raynaud's syndrome, 234
 Reticulum cell sarcoma, 107
 Rodent ulcer, 50
 Ruptured biceps tendon, 334
 Ruptured tendo achillis, 334

S

Salivary ectopic tumour, 129
 Salivary gland, 117
 Salmon pink patch, 31
 Saphena varix, 258
 Sarcoidosis, 108
 Sarcoma
 Retroperitoneal, 307
 Soft tissue, 300
 Scalene syndrome, 239
 Scalene triangle, 239
 Scar, hypertrophic, 45
 Schwannoma, 42
 Schwartz test, 259
 Sebaceous adenoma, 24
 Sebaceous cyst, 22
 Sebaceous horn, 24
 Sialolithiasis, 127
 Sistrunk's operation, 83
 Sjogren's disease, 129
 Spermatocoele, 207
 Spider naevus, 33
 Spina bifida, 281

Spina occulta, 281
 Squamous cell carcinoma, 53
 Sternomastoid tumour, 97
 Strawberry angioma, 32
 Student's elbow, 325
 Sturge-Weber syndrome, 31
 Subhyoid bursal cyst, 84
 Submandibular calculi, 127
 Submandibular gland, 119
 Syringomyelocele, 284

T

Talipes equinovarus, 320
 Tennis elbow, 331
 Testis
 Ectopic, 199
 Retractile, 198
 Undescended, 198
 Thoracic outlet, 238
 Thyroglossal cyst, 80, 81
 Thyroglossal duct, 80
 Thyroglossal fistula, 83
 Torticollis, 97
 Trendelenburg test, 259

Trendelenburg operation, 263
 Trigger finger, 329
 Trosier's sign, 106
 Tuberculous lymphadenitis, 105
 Tumours of the tongue, 145
 Turban tumour, 42

U

Ulcer of the leg, 265
 Arterial, 266, 267
 Bairnsdale, 266, 269
 Cryopathic, 266, 268
 Gummatous, 266, 268
 Infective, 266
 Marjolin's ulcer, 56
 Martorell's, 267
 Neoplastic, 266
 Neuropathic, 266, 268
 Traumatic, 267
 Venous, 265, 267
 Ulcer of the tongue, 145, 151, 152
 Ulnar nerve paralysis – tests, 241
 Umbilical adenoma, 160
 Umbilical calculus, 161

Umbilical carcinoma secondary, 162
 Umbilical endometrioma, 160
 Umbilical fistula, 159
 Umbilical granuloma, 159
 Umbilical hernia, 188
 Urachal cyst, 159

V

Varicocele, 213
 Varicose aneurysm, 251
 Varicose veins, 254
 Ventral hernia, 183
 Virchow's gland, 103
 Vitello-intestinal duct, 157
 Volkmann's ischaemic contracture,
 333

W

Wardill's operation, 292
 Warts, 17
 Warthin's tumour, 121
 Weaver's bottom, 325
 Wryneck congenital, 97

Short Cases in Surgery, 6th edition is, in its present form, a new edition and not merely a reprint. The purpose of this new edition remains the same as those of the previous editions. The entire text has been thoroughly revised and updated. Two new chapters, soft tissue sarcoma, and intestinal stomas have been included. The aim of this book is to help the students, who are preparing for qualifying examinations, in readily grasping the problems of surgery—the problems they have to tackle in their examinations as well as in the wider world outside the examination halls.

It is really gratifying for an author of a book to find it running through many editions. It indicates that the book has proved to be satisfying the needs of the students (and, as they say, also of the teachers) in the institutions of medical learning.

Important features of the book are precisely as follows:

- Most of the surgical problems which are very often posed as 'Short Cases' in examinations have been adequately discussed.
- Each problem has been explained with arguments from all the three dimensions of medical knowledge—anatomy, physiology and pathology. Also the principles of surgical operations have been briefly laid down, wherever possible.
- Discussion has been, whenever necessary, amply illustrated with schematic diagrams to facilitate easy and ready understanding of the subjects.
- Precision is the hallmark of this book. While no necessary information or explanation has been left out, the subjects have been presented in a neat and pointed fashion.
- The book provides necessary aids for ready recapitulation of the important features of problems and their solutions.

Dr. S.K. Bhattacharya, M.S. (Cal.), F.R.C.S. (Edin.), is a Consultant General, Laparoscopic Surgeon and Urologist practising in Calcutta. He worked as Demonstrator in Anatomy at Calcutta National Medical College and Hospital and as Clinical Tutor in Urology, Dept. of Surgery at the same medical college. He also worked as Consultant Surgeon at different district hospitals in West Bengal and retired as Senior Surgeon from Sambhunath Pandit Hospital at Calcutta. He has been a popular teacher during his career in the teaching profession. Dr. Bhattacharya keeps abreast of new developments in the field of surgery.



Book Review

[**Short Cases in Surgery, 4th ed.**] ... this is a superb book ... indispensable to medical students both undergraduates and postgraduates who are preparing for their examination and also who are developing their career in the area of surgery. The charm of this book lies in its simplicity, directness, schematic diagrams. One can read it quickly, easily and with pleasure, ... for practical examination it is presented in the exact way a candidate is desired to answer.

S.K. Mallick, JIMA, Volume 93, April 1995

[**Short Cases in Surgery, 1st ed.**] This book is a welcome edition to the spectrum of examination-oriented clinical books on surgery. ... The diagnostic criteria, classifications, anatomical, physiological and pathological basis of the different congenital and acquired conditions have been described lucidly in a precise and concise manner ... the exact way a candidate for examination is desired to answer. The principles of management and outline of operative procedures will prove to be helpful to the students. ... This will not only be useful to both undergraduate and postgraduate students of surgery but will also be a useful handbook for a busy practising doctor.

Amit C. Ganguli, JIMA, Volume 71, November, 1978



CBS Publishers & Distributors Pvt Ltd

4819/XI, 24 Ansari Road, Daryaganj, New Delhi - 110 002 (India)

E-mail: cbspubs@airtelmail.in; delhi@cbspd.com • www.cbspd.com

New Delhi • Bengaluru • Pune • Kochi • Chennai



ISBN: 978-81-239-2131-0